

**PSYCHOSOCIAL OUTCOMES OF BONE MARROW  
TRANSPLANT FOR INDIVIDUALS AFFECTED BY MPS  
I HURLER DISEASE**

A Thesis submitted for the degree of Doctor of Philosophy

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## **Abstract**

**RATIONALE:** A theoretical model was used to examine the impact of risk and resistance factors on the psychosocial adjustment of children and young people affected by Mucopolysaccharidosis Type I Hurler Disease post-bone marrow transplant. **METHOD:** A sequential exploratory mixed methods design was employed and the study carried out in two phases. In the initial phase qualitative methods were employed in an in-depth study of a small sample of parents of individuals affected by this condition (n=10). They were administered semi-structured interviews, which were analysed using Interpretative Phenomenological Analysis. The themes highlighted were used to inform the design of the second phase, which employed quantitative methods. In this phase forty-four families affected by MPS IH post-BMT participated (44 mothers, 36 fathers, 44 patient participants). This comprised almost the entire population of this patient group in the UK, whose ages ranged from 16 months to 25 years. A face-to-face survey method was used with the mothers, telephone-survey with the fathers and psychometric testing of the patient participants. The measures included risk factors (e.g. physical, cognitive, and adaptive functioning), resistance factors (intrapersonal, stress processing, and social-ecological), and adjustment for both parent and patient participants. **RESULTS:** Data from the qualitative phase revealed a number of themes, highlighting numerous issues pertinent to parents of children affected by this condition. These included perceptions of child vulnerability, uncertainty, perceptions of stress and sources of support, and feeling that others do not understand their situation. Data from the second phase illustrate how stress processing and social-ecological factors make significant contributions to parent adjustment. They also illustrate how intrapersonal, social-ecological, and maternal stress processing factors contribute significantly to patient adjustment. A theoretical path of the determinants of patient psychosocial outcomes was created to illustrate these relationships. **CONCLUSION:** The findings are indicative of how resistance factors moderate the effects that disease- and disability-related risk factors have on parent and patient adjustment as predicted by the model. The dynamic interplay between disease-related risk factors and intrapersonal, social-ecological, and stress processing factors are discussed in relation to parent and patient adjustment outcomes, as are the implications these relationships pose for patient and family support.

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## Abbreviations

MPS	Mucopolysaccharide
MPS IH	Mucopolysaccharidosis Type I Hurler Disease
MPS IS	Mucopolysaccharidosis Type I Scheie
MPS IH-S	Mucopolysaccharidosis Type I Hurler-Scheie
BMT	Bone Marrow Transplant
GAG	Glycosaminoglycans
CNS	Central Nervous System
GvHD	Graft vs Host Disease
ERT	Enzyme Replacement Therapy
MDI	Mental Development Indices
CF	Cystic Fibrosis
VABS	Vineland Adaptive Behaviour Scales
BASC	Behaviour Assessment System for Children
GHQ	General Health Questionnaire
CHQ	Child Health Questionnaire
TBAQ	Toddler Behaviour Assessment Questionnaire
FES	Family Environment Scale
SOC	Sense of Coherence
PSI	Parenting Stress Index
CAPI	Child Abuse Potential Inventory
SIP	Self Image Profiles
ARICD	Association for Research in Infant and Child Development
WISC	Wechsler Intelligence Scale for Children
WAIS	Wechsler Intelligence Scale for Adults

# **CHAPTER ONE**

## **INTRODUCTION**

Mucopolysaccharidosis Type I (MPS I) is a rare chronic childhood condition. It is an inherited disorder of metabolism, which is progressive and in its severest form (MPS IH – Hurler Disease) fatal in childhood. Bone marrow transplant (BMT) has been used to treat this form of the condition since the late seventies in the UK with much success. However, the treatment has its limitations, and while many clinical aspects of the disease are rectified or ameliorated, skeletal abnormalities continue to progress and cognitive and developmental outcomes vary widely. Children growing up with MPS IH post-BMT therefore experience progressive physical disability and their adaptive functioning is challenged. BMT has a potentially long-term survival rate however, and increasing numbers of individuals are living into adulthood with this condition. As a result of BMT having a limited impact on the progression of some aspects of the condition research into MPS I has mainly focussed on medical concerns. Quality of life is however, an issue that has not yet been addressed with this patient group.

No research has explored the impact MPS IH has on the family and consequently the issues facing this group are poorly understood. Given the dearth of research and the rarity of MPS IH, studies that focus on the experiences of individuals and families affected by the condition are important in developing an understanding of the associated psychosocial issues. In turn, an understanding of these issues is important to the development of psychosocial interventions, which are particularly pertinent to those affected by a condition that is disabling, life-limiting, and to date incurable. Having increased the longevity of this patient group, it is now essential that their quality of life be assessed and factors associated with optimal well-being identified. This project therefore aims to assess the quality of life of individuals affected by MPS IH post-BMT by gaining an understanding of their psychosocial functioning and how it is associated with the individual, the condition, and family characteristics.

In order identify the factors that might put a child affected by MPS IH post-BMT at risk of adjustment difficulties, as well as the factors that might protect against risk,

this thesis will examine literature that explores child and family adaptation to living with a variety of childhood chronic conditions. Given the wealth of research evidence that links individual, parent, and family variables with the psychological functioning of children with chronic illness, it is of critical interest that adaptation to MPS IH post-BMT be explored from a family systems perspective (e.g. Seligman and Darling, 1997) and considers the factors that might contribute to and protect against adjustment difficulties. Before evaluating the psychosocial factors associated with adaptation to childhood chronic illness however, an account of the natural history of MPS IH will be given. This will be followed by some discussion on the diagnosis of this condition, and the physical and neurological outcomes of bone marrow transplant. The literature review will continue with a critical exploration of the psychological impact that paediatric bone marrow transplant can have on a child and their family. The rationale for this is to provide the reader with an appreciation of the severity of the condition, the high-risk, traumatic, and uncertain nature of the treatment, and the psychological sequelae of BMT for the whole family. Simultaneously, the biomedical aspect of a stress and coping model for MPS IH post-BMT can be set in context.

## **1.0 Mucopolysaccharidosis Type I – Disease Classification and its Natural Progression**

“The mucopolysaccharidoses are a group of inherited lysosomal storage disorders with distinctive phenotypes and a progressive course” (Colville and Bax, 1996:31). There are seven recognisable sub-types of MPS, as shown below, and there are a further thirteen related diseases.

MPS I	Hurler (MPS IH), Hurler Scheie (MPS IH-S), and Scheie (MPS IS);
MPS II	Hunter;
MPS III	Sanfilippo;
MPS IV	Morquio;
MPS VI	Maroteaux Lamy;
MPS VII	Sly;
MPS IX	Hyaluronidase Deficiency

(MPS V was originally called Scheie disease, but has been reclassified as MPS I after it was discovered to be the result of the same enzyme deficiency. MPS VIII has also been reclassified).

Although all of the MPS diseases are progressive in nature, the severity and speed of the disease progression can vary. Each phenotype has its own continuum of severity, and vital organs, the skeleton, joints, and the central nervous system are affected in differing degrees. However, all of the mucopolysaccharidoses are inherited disorders of metabolism and are characterised biochemically by a deficiency of specific lysosomal enzymes. MPS I is characterised by the deficiency of the lysosomal enzyme  $\alpha$ -L-iduronidase, which is needed to degrade the glycosaminoglycans (GAGS) dermatan sulphate and heparan sulphate. As a multi-systemic disorder, in its severest form (MPS IH - Hurler disease), all tissues in the body have some degree of GAG storage and disease. As the build-up of GAG increases over time, the child affected by MPS IH will develop physical features, which are characteristic of the disorder. These include a large head with a prominent forehead, short neck, broad, flat nose with wide upturned nostrils, enlarged tongue and thickened lips, widely spaced teeth, broad gum ridges, and coarse hair. Children with MPS IH also have distended abdomens due to enlargement of the liver and spleen (hepatosplenomegaly), they have a characteristic way of walking and holding their arms due to joint contractures at the shoulders, elbows, hips, and knees (dysostosis multiplex), and their hands are clawed. They are prone to carpal tunnel syndrome, and inguinal and umbilical herniae are common. Further complications include cardiac disease, pulmonary disease, hydrocephalus, spinal deformity (kyphosis/scoliosis), problems with hearing and eyesight and progressive mental retardation. Children with MPS IH may grow relatively fast in their first year of life, but growth then slows and ultimately stops around the age of three years. The average height achieved by a child affected by MPS IH is approximately 100cms. With regard to intellectual ability, a similar pattern of development and decline can be seen. Without intervention children with classic Hurler disease usually die before their tenth birthday.

Although MPS I has three different disease classifications under its umbrella, they are all characterised by the deficiency of the same lysosomal enzyme, and are therefore

part of a spectrum of the same disorder. Scheie disease is the more attenuated form of MPS I and is often referred to as the 'mild' form (MPS IS). However, although individuals affected by Scheie disease do not usually experience learning difficulties, and grow to a relatively normal height, they are still affected by a progressive disease and can experience physical problems similar to those affected by Hurler disease. These are considered to be less progressive however, and individuals affected by Scheie disease can expect a relatively normal life span. There are other individuals whose disease pattern falls between the two ends of the spectrum, and this form of MPS I is referred to as Hurler-Scheie disease (MPS IH-S).

## **1.1. Diagnosis**

MPS I is an autosomal recessive lysosomal storage disorder. In other words, in order to give birth to a child with MPS I, faulty genetic mutations are required from both parents. It is therefore, a rare condition, the incidence of which has been estimated at 1 per 100,000 (Spranger, 1972; Lowry and Renwick, 1978). In order to develop the most severe phenotype, Hurler disease, two fully deleterious mutations need to be bound. The most common of these genetic mutations have been identified as W402X and Q70X. Late onset or milder forms of the disorder are the result of combinations of mutations that leave enzyme function partially intact. These combinations of mutations are less common as they are the result of the binding of a wider range of genetic mutations. Although such genetic mutations are recognised as the cause of MPS I, DNA testing is not the usual route to diagnosis. Since the condition is extremely rare, diagnosis through pre-natal genetic counselling is unlikely, unless its heredity has already been established in the family. Diagnosis is therefore made after the emergence of early signs and symptoms. However, since these symptoms are commonly seen in many young children, and since many general practitioners are not specialists in lysosomal storage disorders, early diagnosis can be problematic.

A study by Cleary and Wraith (1995) describes the presenting features of MPS IH, and illustrates how the non-specific nature of certain early symptoms, such as recurrent upper respiratory tract infection, did not alert medical professionals to the more serious underlying problem. In Cleary and Wraith's study the average age of

diagnosis was 9.4 months, with an age range of 3–18 months. Of the main presenting features leading to investigation and diagnosis, rhinitis and recurrent inguinal herniae were the earliest. However, at six months of age, skeletal abnormalities became a common presentation. These usually presented themselves as a gibbus formation, an asymmetrical chest, prominent sternum, and bulging forehead. At six months of age recurrent upper respiratory infections and herniae continued to be common complaints. After nine months of age the facial features that are characteristic of children affected by Hurler disease became obvious in several of the children and this alone led to diagnosis. Also at nine months, failure to pass the health visitor's hearing test was frequently reported. From 12 months onwards, the skeletal abnormalities and characteristic facial features typically associated with MPS IH became recognisable in most of the children in the study. If not diagnosed prior to this stage, diagnosis followed soon after these developments.

In recent years more awareness has been raised amongst physicians, regarding lysosomal storage disorders. It is considered that certain clinical features should lead to suspicion early on and prompt screening procedures. Some features that should lead to suspicion include hepatosplenomegaly, coarse facial features, an unusually large head circumference, skeletal dysplasia, corneal clouding, and developmental delay. Once the early signs of MPS IH have been recognised, two methods of diagnosis are commonly used. The first is the measurement of abnormally high urinary GAG levels, and the second is by testing the level of enzyme activity in a blood or skin sample.

Since MPS I is a progressive disorder it is imperative that diagnosis is established as early as possible, as although there is no known cure, treatments and therapies can be used to manage the disease. Clinical trials of enzyme replacement therapy (ERT) have been completed with great success, and the treatment was licensed and made available in the UK in 2004. This is a genetically engineered  $\alpha$ -L-iduronidase, known as Laronidase (Wraith, 2005; Wraith, Hopwood, Fuller, Meikle et al., 2005; Kakkis, McEntee, Schmidtchen, Neufeld et al., 1996; Shull, Kakkis, McEntee, Kania et al., 1994). Gene replacement and substrate reduction therapies are being actively researched, and bone marrow transplantation is commonly practiced. However, the efficacy of these therapies depends upon early diagnosis and treatment, before the

onset of irreversible pathology. It also depends upon the severity of the condition: the phenotype. For example, although ERT has proved to be effective in improving many of the clinical features of MPS I, it does not impact on the central nervous system (CNS), and so is more suitable for the treatment of the milder or more attenuated forms of the disorder. ERT is being used however, to treat infants affected by MPS IH in the short-term, pre- and post-BMT, to improve the child's physical health status in preparation for transplantation (Grewal, Wynn, Abdenur, Burton et al., 2005). The longer-term benefits of this treatment, if any, will not be realised for many years.

## **1.2. Bone Marrow Transplantation as Treatment for MPS IH**

Bone marrow transplantation has provided a method of correcting enzymatic deficiencies in several lysosomal disorders. Hobbs, Hugh-Jones, Barrett, Byrom, et al. (1981) reported that allogeneic BMT could dramatically improve the somatic features of MPS IH. Following BMT, they observed an increase to donor level in the deficient enzyme in a one-year old child. They also observed the normalisation of urinary GAG and decreased hepatosplenomegaly. Since this original report, hundreds of children with storage diseases have received allogeneic BMTs worldwide, including many with MPS IH. Matched sibling donor BMT has resulted in metabolic correction. Accumulated substrates are found to decrease to normal levels, and many clinical features such as hepatosplenomegaly (Resnick, Krivit, Snover, Kersey et al., 1992), sleep apnoea (Malone, Whitley, Duvall, Belani et al., 1988), cardiac failure, and pulmonary disease (Whitley, Belani, Chang, Summers et al., 1993) are either resolved or ameliorated. Correction of odontoid dysplasia (dysplasia of the first two vertebrae at the top of the spine) can also be achieved, which uncorrected has the potential for life threatening cervical spinal cord injury (Hite, Peters and Krivit, 2000; Tandon, Williamson, Cowie and Wraith, 1996).

However, BMT is not a perfect treatment. Despite stable engraftment, skeletal abnormalities continue to progress, necessitating surgical interventions. Furthermore, with regard to neuropsychological capabilities, BMT outcomes vary widely. If transplantation takes place early in life, before irreversible neuropsychological damage occurs, a rate of learning with low normal intelligence can be achieved

(Peters, Balthazor, Shapiro, King et al., 1996). Escolar and Kutzberg (2002) illustrate the importance of early transplantation in relation to mental development. They found that if transplanted under 12 months, mental development indices remained in the low average range at two years of age, remaining stable up to three years after transplant. However, if transplanted between 12 months and two years of age, mental development was found to be moderately delayed by 4-5 years of age. Thus, neurodevelopmental outcomes show significant improvement if BMT is carried out early in infancy. As a result of their findings, newborn screening programmes are being advocated. It is hoped that, if infants are treated pre-symptomatically, it may be possible to prevent learning difficulties for children with MPS IH.

While longer term outcomes of BMT for MPS IH remain to be uncertain and BMT itself a high risk procedure, research into BMT for this condition shows encouraging results in terms of effectiveness and survival. Effectiveness of BMT is defined as “engrafted survival with continuing cognitive development” (Peters, Shapiro, Anderson, Henslee-Downey et al., 1998). Since the late 1980s physicians at the Debrousse Hospital in France have been transplanting infant patients affected by MPS IH (Souillet, Guffon, Maire, Pujol et al., 2003). In that time they have performed a total of thirty haematopoietic stem cell transplants on twenty-seven children. Four of the primary transplants failed to engraft, two of these patients went on to have a second transplant, one resulting in successful engraftment. Thus three of the twenty-seven transplants failed to engraft. Two of these patients went on to experience disease progression and at approximately 4-5 years follow-up, one had died due to disease progression. Three patients who successfully engrafted died as a result of infection. At follow-up, twenty-one (78%) of the twenty-seven patients had functional grafts with favourable long-term outcome.

Figures from the US show that between 1983 and 1995, of fifty-four MPS IH patients underwent BMT, thirty-nine (72%) engrafted after the first transplant (Peters et al., 1998); and more recent figures show an 85% event-free survival rate at a median of 905 days after transplantation of unrelated cord-blood stem cells (Staba, Escolar, Poe, Kim et al., 2004). At present there are no recent published data regarding BMT engraftment and survival rates from the UK. However, although it is difficult to give estimations of long-term survival for MPS IH patients, it is believed that with bone



marrow or cord blood transplants, the likelihood of long-term survival is as high as 80% (Peters, Orchard, DeFor, Grewal et al., 2001; Staba, Martin, Ciocca, Allison-Thacker et al., 2001).

With regard to cognitive function, as previously discussed, patients transplanted before 24 months of age with a baseline mental development indices (MDI) greater than 70 can achieve improved cognitive function and favourable outcome post-BMT (Peters et al., 1996; 1998). However, more recent figures have found children transplanted before 12 months of age to be able to achieve MDI within the normal range (Escolar and Kutzberg, 2002); and with cord blood transplants, at 66 months of age, mean scores of cognitive function have been found to fall within the normal range post-transplant (Staba et al., 2004). Thus, in terms of cognitive functional outcomes early transplantation is advocated, preferably before 12 months of age (Hopwood, Vellodi, Scott, Morris et al., 1993; Shapiro et al., 1995). In terms of adaptive function however, age at transplant has not been related to later adaptive level (Bjoraker, Delaney, Peters, Krivit et al., 2006), though following transplantation, adaptive functions, including communication, daily living skills, socialisation, and motor functions, have shown continued development albeit at a slower rate than peers. Children with good cognitive function pre- and post-BMT show good adaptive functions however (Bjoraker et al., 2006).

Outcomes of BMT for children affected by MPS IH are therefore extremely varied and dependent upon a variety of factors. These include the progression of the disease before transplantation, successful engraftment and whether a second transplant is required, thus allowing more time for the disease to progress, and BMT-related complications such as infections and Graft-vs-Host disease (GvHD). Research to date suggests that BMT can reduce substrate accumulation, reverse some clinical features of the disease, and can improve neurological function. The treatment also has a long-term survival rate. However, musculoskeletal problems persist and, although patients can remain mobile some years post-BMT, increasing pain and stiffness of the hips and knees, and spinal curvature are a probability (Vellodi, Young, Cooper, Wraith et al., 1997). Essentially, BMT *changes* the aetiology of the condition rather than cures it. It thus creates a unique condition which has uncertain medical outcomes and uncertain

outcomes in terms of physical and adaptive functioning. It thus brings with it a potential array of psychosocial issues for the individual and their family.

### **1.3. Expectations of Bone Marrow Transplant**

Families of children affected by MPS IH often receive an MPS diagnosis for their child after a period of numerous referrals, misdiagnoses, and misinformation. It is a devastating diagnosis, which requires the family to process a wealth of complex medical information and to contemplate the deterioration, disfigurement, and early death of their child. Since early treatment is related to more favourable functional outcomes the family are simultaneously required to make the decision whether or not they wish their child to undergo bone marrow transplantation: a high risk treatment. The diagnosis of a child's chronic illness is an extremely stressful time for parents, and they often fail to fully comprehend their child's diagnosis when it is initially given (Whyte, 1992). This gives rise to an enormous amount of anxiety regarding the child's condition and potential outcomes, which is a major stressor at the time of diagnosis (Cohen, 1993; Miles and Holditch-Davis, 1997). Uncertainty has been described as a cognitive state which results from an event being inadequately defined or classified, and it is purported to be a major factor influencing expectations about treatment and prognosis (Hilton, 1992). It has further been highlighted as 'a major perceptual variable' hindering the psychological management of a condition (Swallow and Jacoby, 2001:70). While the receipt of a diagnosis can provide families with a sense of relief it can also pose another set of uncertainties, particularly when treatment options are high-risk, treatment outcomes uncertain and future quality of life unpredictable (Cohen, 1997). Such uncertainty has been found to elicit a grief response in mothers of children diagnosed with chronic illness (Gibson, 1995).

Further to this, the cognitive processing of emotionally devastating and unfamiliar information can have implications for decision-making. Much of the information that needs to be conveyed about severe chronic conditions is often not concrete and imaginable to lay people and as a result can be difficult to remember (Paivio and Csapo, 1973). Much of the information presented is also threatening in nature, which itself evokes intense emotions. Research has demonstrated how powerful negative

emotions can interfere with memory (Loftus and Burns, 1982). More recently, a shock reaction to the news that one's child has a life-limiting and incurable condition has been found to impede the comprehension and remembering of medical information presented (Jedlicka-Kohler, Gotz and, Eichler, 1996). Thus, with confusion about diagnosis, uncertainty about treatment outcomes and longer term quality of life, parents' ability to maintain realistic expectations about BMT for MPS IH is extremely difficult. This is exacerbated by the progressive nature of the disorder and parents' difficulty imagining the manifestations of the condition when they are not yet apparent.

More immediate concerns about saving the life of their child are therefore likely to take priority over expectations of longer-term outcomes. Parents who choose to proceed with BMT as treatment for MPS IH are doing so to give their child the chance of a life and a future. They do not want their child to die. Longer-term outcomes are therefore perhaps further back in parents' minds at this time. Furthermore, when faced with enormous stress or adversity, unrealistic optimism and denial of threat are often employed as adaptive coping mechanisms (Taylor and Brown, 1988). Individuals commonly employ an 'optimistic-bias' (Weinstein, 1980; 1982) whereby they underestimate the possibility that negative events will happen to them. Research has found that despite the provision of extensive information regarding possible post-BMT physical and psychosocial morbidity, individuals use biases in their interpretation and processing of information which perpetuate optimistic expectations. When faced with the 'life or death' decision to undergo BMT, a variety of factors combine to increase the likelihood that patients will discount or dismiss the possibility that long-term post-BMT complications will occur (Andrykowski, Brady, Greiner, Altmaier et al., 1995). Thus, for parents of children affected by MPS IH, when evaluating their expectations of BMT, it is likely that their focus is on the here and now and on the chance of life for their child, rather than on longer term functional outcomes.

## **1.4. Giving Informed Consent**

With regard to informed consent, some research has explored the BMT decision-making process. Although informed consent is not requested until after the patient has been allowed full consideration of the treatment based on a realistic understanding of the risks and possible outcomes, it is suggested that the decision to go ahead with the treatment is made when bone marrow transplant is first suggested as a treatment option (Andrykowski, 1994). It is posited that 'this early commitment to BMT [is seen as] a function of the life-threatening nature of the patient's illness' (Andrykowski, 1994:359). A study by Kodish, Lantos, Stocking, Singer et al. (1991) describes how many parents of children are not willing to accept the risk of mortality for their child. The study showed that 24% of a sample of mothers of children with sickle cell disease was not prepared to accept any risk of mortality through BMT, however small. Only 37% were willing to accept 15% risk of mortality within 30 days of BMT, with 8% willing to accept at least a 50% risk of mortality within 30 days of BMT. Since the natural course of sickle cell disease is chronic debilitation rather than death, these findings are not necessarily contrary to Andrykowski's view, even though they are purported to be. The ratio of parents of children affected by MPS IH that choose not to undergo BMT to the parents that choose to go ahead with the treatment is not known. However, anecdotal evidence suggests that the decision to go ahead with BMT is made as soon as a suitable bone marrow match is found. This suggests that the decision to go ahead with the transplant is already made, but that the final decision is dependent upon finding a suitable match. Once a match was found, many families reported that the decision to go ahead 'had been made for them'.

Making the decision to allow one's child to undergo bone marrow transplantation is an extremely distressing and traumatic experience. It is an invasive and high-risk procedure, and while for MPS diseases it is a potentially life saving treatment, it is not a curative one. Prior to giving consent parents are provided with critical information regarding the risks and benefits of BMT relative to any alternative treatment. They are also given information regarding possible patient lifestyles after BMT. However, as previously discussed, prognoses are difficult to give with any certainty. Thus, in giving consent, not only do the family have to consider the high risks involved in the

procedure itself, they also have to consider the uncertain outcomes of treatment and the heterogeneous nature of the condition post-BMT, and weight up the risks and benefits against the natural progression of the disease. All of which are difficult concepts to comprehend.

While no research has explored the experiences of families with infants affected by MPS IH undergoing BMT, research of this procedure being used to treat other conditions, such as cancer, has illuminated the psychological impact the treatment process can have on the child and on family members. In particular, mothers have been reported to experience elevated levels of distress throughout the decision-making process (Dermatis and Lesko, 1990) and at the pre-admission stage (Streisand, Rodrigue, Houck, Graham-Pole et al., 2000), to experience persistent depression before transplantation, and to feel doubtful about their decision to submit their child to BMT both before and after the procedure (Pot-Mees and Zeitlin, 1987; Pot-Mees, 1989). Furthermore, children undergoing BMT have been reported as experiencing distress prior to BMT, throughout the process, and on discharge (e.g. Kronenberger, Carter, Stewart, Morrow et al., 1996; McConville, Steichen-Asch, Harris, Neudorf et al., 1990; Phipps, Brenner, Heslop, Krance et al., 1995; Wiley and House, 1988).

## **1.5. Paediatric Bone Marrow Transplant**

‘BMT is an aggressive, high-technology medical procedure associated with an array of psychological and physical stressors including isolation in a germ-free environment, rapid and uncertain fluctuations in medical status, prolonged hospitalisation, frequent invasive medical procedures, treatment-related physical side-effects, extreme dependence on hospital staff, repeated infections, GvHD, and, of course, the possibility of death’ (Andrykowski, 1994:357). With regard to paediatric bone marrow transplantation, as well as having to undergo distressing procedures common to the treatment such as chemotherapy, bone marrow aspirates, and lumbar punctures, children are subject to additional stressors associated with the treatment process and post-process restrictions, such as having to spend time away from family and friends due to lengthy hospitalisations, and having continued restricted social contact on discharge (Streisand et al., 2000). Research indicates BMT to impact on

children psychologically. They are reported as experiencing clinically significant levels of psychological distress at the pre-transplant stage (e.g. Kronenberger et al., 1996). They are also reported as being more stressed, having low social competence, low self-esteem, emotional difficulties, and multiple concerns upon discharge (McConville et al., 1990; Phipps et al., 1995; Wiley and House, 1988). Other research has found children who have undergone BMT to experience post-traumatic stress symptoms three (Stuber, Nader, Yasuda, Pynoos, and Cohen, 1991) and six months post-procedure (Pot-Mees and Zeitlin, 1987; Pot-Mees, 1989).

As well as the individual undergoing the procedure, of course the BMT process can have a tremendous psychological impact on other family members and can impact on the whole family and the way that it functions. Available research has demonstrated how other family members can experience psychological distress when a child family member undergoes BMT. Feelings of rejection have been reported amongst non-donor siblings (Pot-Mees and Zeitlin, 1987; Pot-Mees, 1989) and they have been reported as showing more problems in school than donor siblings (Packman, 1999). Feelings of anxiety concerning the transplant have been reported amongst donor siblings (Pot-Mees and Zeitlin, 1987; Pot-Mees, 1989; Packman, 1999) and they have been shown to have lower self-esteem than non-donors (Packman, 1999). Siblings are also reported as experiencing temporary behavioural problems, such as enuresis, disobedience, and sleep and eating disturbances. These disturbances are ostensibly more common amongst sibling donors. However, the psychological reaction to BMT of both donor and non-donor siblings is suggested to include some post-traumatic stress symptomology (Packman, 1999). Marital relationships can also be put under stress as a result of prolonged separation. Mothers often experience extreme anxiety and depression as already discussed. They are also reported as suffering sleep disturbance, and mental and physical exhaustion, as a result of staying in the hospital with their child. Subsequently, fathers are reported as feeling 'out of touch', which has been suggested as contributing to them harbouring unrealistic fears about their child's condition (Pot-Mees and Zeitlin, 1987; Pot-Mees, 1989).

Suffice to say, paediatric BMT is an arduous process that involves emotional extremes. It is also a lengthy experience, which has been conceptualised by Pfefferbaum, Lindamood and Wiley (1978) as a ten-stage process. The authors

observed 30 child sufferers of leukaemia and aplastic anaemia and their families in a medical centre throughout the bone marrow transplant procedure and outlined a number of psychological responses. The children's ages ranged from five to nineteen years and they were admitted to hospital between 1973 and 1977, when BMT was a relatively new treatment. This viewpoint is a useful one to consider as not only does it provide an insight into the psychological reactions of children and families undergoing this aggressive procedure, it does so at a time when BMT was a new treatment so highlights the uncertainty experienced by families and their tendency to have misconceptions and false expectations. Shortly after this time BMT started to be used to treat other conditions including MPS IH.

Stage Zero is the first of the stages proposed, where the parents are *introduced to the concept* of BMT and offered it as a treatment option. This is suggested as being a time of false expectations and misconceptions about the treatment. However, families are given the opportunity to meet with transplant patients and their families during clinic visits. Stage One, *the preadmission experience*, is the point where the family are required to give informed consent to their child's bone marrow transplant based on a realistic understanding of the risks and possible outcomes of the treatment. Once informed consent has been given, the child and family enter Stage Two, which is a time of initial *inpatient evaluation*. During this time the psychosocial aspects of the procedure are considered and how both the child and their family react to the stresses associated with the experience are examined and monitored. Once cleared as physically and psychologically fit for transplant, the child is put into *isolation*, which is Stage Three of the BMT process. During this stage the authors report that young children are not particularly disturbed by the experience as their world in the form of school work and toys can be brought to them. Stage Four is *day minus one*. This is purported to be one of the most significant days, as the donor is admitted to hospital and the sick child is administered total body radiation or chemotherapy in more modern procedures. This is particularly harrowing for both the child and the family as the treatment renders the child sick with 'shaking chills, nausea and vomiting, and diarrhea' (Pfefferbaum et al., 1978:626). The authors report parents' feelings of separation and helplessness to be particularly intense at this time.

The bone marrow transplant itself is the following stage, *transplant day*. However, it is viewed as anti-climactic by the family, in the wake of the trauma of the total body radiation. Although it is the most significant stage, it is perhaps the calm before the storm, as it is shortly followed by an anxious *waiting period*. During this stage, the child is kept in isolation and, although parents feel more confident in the day-to-day management of their child's care, it is a time of uncertainty as they wait to see if the bone marrow has taken. *The take* is the seventh stage of the process, which takes place two weeks after the transplant, and is when the patient's marrow shows activity. The authors report that children can attribute the cause and course of the illness to themselves, and that they can feel responsible for the success or failure of the treatment. In the case of families affected by MPS IH, a further *wait* is in store to see if enzyme is being produced and to what extent. Even when the news is good and the transplant has been a success, the family are now at a critical stage, as the child is at high risk for bleeding, infection, and graft vs host disease (GvHD), when the immune cells in the transplanted bone marrow reject the host body. The child therefore has to continue to be kept in isolation. During this stage children are reported as exhibiting unsociable behaviour, with some becoming depressed and withdrawn. They can also act-out and behave negatively towards parents and hospital staff. *Preparation for discharge* is the ninth stage. During this stage the child is required to wear a mask as protection from known pathogens. They also appear weak and thin and have often lost their hair. They are thus reported as feeling insecure regarding their appearance and fear rejection. Parents equally feel overwhelmed by their child's fragility and have difficulty coping with their child still being significantly unwell when they were supposed to be 'well'. *After discharge* from the hospital, the tenth stage of the process, the child is required to stay in semi-isolation in the home for up to six months, when infections and GvHD are still a possibility.

These observations are useful as they provide an insight into the psychological impact the BMT process can have on child recipients and their families, and of the fragility of the child following this aggressive procedure. As with the majority of studies that have explored paediatric BMT and its impact on the child and family however, the children observed were five years of age and over. Little research has studied the experiences of families with very young infants undergoing BMT and none has explored the experiences of families affected by MPS and related diseases. Evidently



the psychological impact on the child would entail fewer conscious perceptions in early infancy, but the experience for the parents would ostensibly not differ. However, as previously mentioned, BMT is not a curative treatment for MPS IH as it is for leukaemia and there are many uncertainties surrounding outcomes. Some aspects of the condition continue to progress and little is known about longevity, patient functioning, and quality of life. The concept of the fragile child post-BMT is of particular interest then and is a key component of the research being undertaken here. For many families with children affected by MPS IH it is possible that perceptions of child vulnerability prevail beyond discharge from hospital and the subsequent period of semi-isolation, which could impact on parents' psychological management of the condition (Swallow and Jacoby, 2001), their adaptation, the functioning of the family, the way they parent their child, and ultimately the psychosocial functioning of child. These issues will be discussed in more depth in the next chapter.

## **1.6. Conclusion**

MPS I Hurler Disease is a devastating progressive condition which is genetically transmitted and untreated will result in death within the first decade of life. It is routinely treated with bone marrow transplant in early infancy. However, the outcomes of the treatment are uncertain as it is non-curative and some aspects of the disease continue to progress. Research has addressed a number of issues related to treatment limitations, treatment options, orthopaedic issues, adaptive and cognitive functioning, and other medically related topics. However, the psychosocial functioning of children and young people affected by this condition post-BMT has not been studied to date. Post-BMT, MPS IH continues to be a severe, disabling, and chronic condition, which presents with short stature, distinctive facial features, an awkward gait, spinal curvature, auditory and visual impairment, and progressive physical disability. Affected children also have some degree of learning disability, which ranges from profound to within the normal range, and adaptive functioning is challenged. Furthermore, management of the bone and joint disease that continues to progress can require significant and sometimes repeated orthopaedic surgery, and this together with the medical management of the condition in general can result in repeated hospitalisations and time away from the home and school. Children and

young people growing up with this condition therefore face a multitude of issues that may impact on them psychosocially.

Equally, for parents, the experience of having a child with a chronic condition can present with an array of stressors that can make adaptation difficult. Having one's infant diagnosed with a life-limiting condition such as MPS IH is a devastating experience. Furthermore, seeing that child undergo the traumatic, high-risk, and uncertain treatment of bone marrow transplant is tremendously distressing. Moreover, for parents of children affected by MPS IH, uncertainty prevails, particularly in terms of life expectancy before, during, and after BMT, functional and health outcomes of BMT, and outcomes of treatments such as orthopaedic surgery. Parents also have little idea of what to expect in terms of their children's future quality of life post-BMT, however long or short that life may be. The impact that these and other factors may have on parent adjustment are crucial to our understanding of children's psychosocial functioning post-BMT, as a wealth of research evidence exists, which links individual, parent, and family variables with the psychological functioning of children with chronic illness. While there are many aspects of the condition itself that can impact on the child psychosocially, the serious, life-limiting, and uncertain nature of the condition and its treatment have additional implications for parent adjustment and family functioning, and in turn for child psychosocial outcomes.

While medical research has given children affected by MPS IH the chance to live longer lives and continues to try to improve biomedical outcomes post-BMT, the psychosocial outcomes of BMT have yet to be explored. The fact that this area of study has been neglected with this patient group is a concern that has been raised by parents of MPS-affected children and by the patient groups that support them and their children, and it is this concern that has led to this research being carried out. While strivings to perfect treatment regimens continue to be important, it is also now imperative that some attention is drawn to the quality of individuals' lives post-BMT and to the factors that contribute to it, so that patient outcomes can continue to be improved upon. It is the aim of this research therefore, to assess the quality of life of individuals affected by MPS IH post-BMT by gaining an understanding of their psychosocial functioning and how it is associated with the individual, the condition, and family characteristics. It is hoped that this will illuminate factors that might

facilitate individual and family adaptation. This in turn will go some way to honing future research of this nature and to developing appropriate intervention strategies that may help individuals and their families adapt to living with this condition.

The following chapter will examine literature in the field of health psychology that explores family and individual adaptation to chronic illness. The purpose of this is twofold. The literature will first be explored in order to identify a model of adaptation that appropriately conceptualises children's adaptation to MPS IH post-BMT and can be applied empirically to the study of the affected individuals' psychosocial functioning post-BMT. Secondly, the literature will be explored in order to identify factors that may be pertinent to the psychosocial functioning and adaptation of individuals and their families affected by MPS IH post-BMT. Since no research has explored the psychosocial outcomes of BMT for individuals affected by MPS IH it will be necessary to draw on work that has explored other chronic and disabling childhood conditions, including spina bifida, cystic fibrosis, juvenile rheumatoid arthritis, sickle cell disease, and diabetes.

## **CHAPTER TWO**

### **LITERATURE REVIEW**

Having established the severity of MPS IH, its causes, diagnosis, treatment, and the implications the BMT process and outcomes have for child patients and their families, this chapter reviews theoretical models and prior research exploring child and family adaptation to chronic illness.

#### **2.0. Models of Adaptation to Chronic Illness**

It is estimated that 31% of children are affected by one or more chronic conditions (Newacheck, 1994). Chronic illness is described as ‘a disorder with a protracted course which can be progressive or fatal, or associated with a relatively normal life span despite impaired physical or mental functioning. Such a disease frequently shows periods of acute exacerbations requiring intensive medical attention’ (Mattsson, 1972). As well as the child however, such conditions affect a far wider network of individuals, including family members and friends, and the wider community. This is not least due to families increasingly becoming the primary caregivers when a child is ill (Stewart, Ritchie, McGrath, Thompson and Bruce, 1994). Research has demonstrated that there is a wide variation in psychological adjustment to chronic illness; while some individuals and families experience adjustment problems, others adapt in successful and resourceful ways (e.g. Venters, 1981; Sawyer, 1992). A systems approach to childhood chronic illness and disability (e.g. Seligman and Darling, 1997) is therefore important in appreciating the dynamic nature of family functioning and how this is affected by the difficulties experienced by one individual. It is well documented that family functioning is recognised as an important contributor to both child (e.g. Drotar, 1997; Quittner and Digirolamo, 1998) and parent adjustment (e.g. Moos and Moos, 2002; Warfield, Krauss, Hauser-Cram, Upshur et al., 1999). It is also reported that the ability to cope with living with a chronic condition is not directly related to the severity or the nature of the condition (e.g. Beresford, 1994), but to a number of other factors that are involved in the development of adjustment problems (Wallander and Varni, 1998).

A number of models of adaptation to chronic illness have been developed which conceptualise illness as a potential stressor and view adaptation from a systems-theory perspective. These have been extremely influential in shaping subsequent conceptualisations and in developing intervention initiatives to enhance adaptation. The role of psychological processes in children's adjustment to chronic illness was first put forward in a model formulated by Pless and Pinkerton (1975). The model viewed adjustment as being multiply determined by transactions between the individual and his or her environment. The model particularly highlighted the role that the family and social environment play in shaping the child's intrinsic attributes such as temperament and personality and his or her response to the illness, which in turn shape the psychological processes, namely self-concept and coping style, that are seen as key to psychological adjustment to chronic illness. The model further proposed that the functioning of the child effects the way others respond to the child, which in turn reciprocally has an effect on the child's future functioning. Thus, adjustment is seen as changing over time and psychological functioning reflective of the nature and cumulative effect of earlier transactions.

Pless and Pinkerton's (1975) model of adaptation proved to be seminal in the field of chronic illness (Thompson and Gustafson, 1996) as three conceptual components have been included and built upon in numerous subsequent models. These are the conceptualisation of illness as a stressor, the idea that adaptation is an ongoing process, and the emphasis on coping methods being important to the process of adaptation. At the same time as Pless and Pinkerton's work with chronic illness, conceptualisations of stress and coping in both the psychological and medical literature were making great strides. Stress was broadly defined as any situation 'in which environmental demands, internal demands, or both tax or exceed the adaptive responses of an individual, social system, or tissue system' (Monat and Lazarus, 1977:3). Thus, whatever the type of stress (physiological, psychological, or social (Monat and Lazarus, 1977), it was defined as such if it exceeded an individual's ability to cope with it. However, it was recognised that individuals differed in their responses to stress and differentially perceived situations as stressful. The role of cognitive processes, including appraisals, expectations of efficacy and control, and causal attributions were then emphasised in psychological approaches to

understanding the variability in the experience of stress (Thompson and Gustafson, 1996).

The transactional model of stress and coping as proposed by Lazarus and Folkman (1984) conceptualised illness as a stressor and responses to the stressor as mediated by appraisals of the situation and coping processes. The meaning that the illness holds for the individual is paramount to adaptation in this instance. Similarly, Leventhal and colleagues (e.g. 1997, 2003) proposed their common sense model of self-regulation of health and illness (CSM), which highlighted coping as a mediating factor. They conceptualised coping processes and appraisals of coping strategies as fundamental to adaptation as they ostensibly influence illness representations, or the way in which an individual makes sense of their illness and its symptoms. Coping processes and appraisals thus have an impact on future coping efforts and ultimately on the way one feels about him or herself and their situation. The CSM has been used extensively to explore adaptation to a number of chronic conditions including musculoskeletal disease (Hill, Dziedzic, Thomas, Baker and Croft, 2007), chronic fatigue syndrome (Moss-Morris, Petrie, and Weinman, 1996), multiple sclerosis (Vaughan, Morrison and Miller, 2003), and rheumatoid arthritis (Scharloo, Kaptein, Weinman, Hazes, Breedveld, and Rooijmans, 1999; Treharne, Lyons, Booth, and Kitas, 2005). This has been done via the development of the Illness Perception Questionnaire (IPQ) and its revised version, the IPQ-R (Weinman, Petrie, Moss-Morris, and Horne, 1996; Moss-Morris, Weinman, Petrie, Cameron, and Buick, 2002). It has also been used to explore a number of paediatric conditions including type I diabetes (e.g. Urquhart Law, 2002). However, while both approaches consider internal and external coping resources and contextual factors that might influence cognitive processes, and consider the process of adaptation as ongoing, 'specific predictions about the role of and interaction with significant others' (Hale et al., 2007:905) are not made. Thus these models fail to consider the direct and indirect effect relationships with family members, friends, and members of the wider community might have on adjustment, including illness representations, for the individual and their parents.

A number of models of family functioning have been developed in the field of chronic illness, which examine in particular how families adapt, cope, and provide care. The T-double ABCX model of family adjustment and adaptation (McCubbin and

Thompson, 1987) has been widely applied to families. This model further expands on McCubbin and Patterson's (1981) extension of Hill's classic family stress theory and ABCX family crisis model (1958). While maintaining the key components of the originally extended model: 'pile-up' of stressors (A), family resources to meet the crisis (B), family's interpretation of the crisis event (C), and the family crisis itself (X), this model underscores the importance of family functioning patterns for adjustment and adaptation. Further conceptualisations of this model have examined the relationship between family functioning variables over time and consider family adaptation as a dynamic and ongoing process which changes as the family moves through the life cycle (McCubbin and McCubbin, 1991). Resilience has also been emphasised in the adaptation process (McCubbin and McCubbin, 1993; 1996). These theoretical bases are helpful in illustrating how families function as a whole to handle demands, and how supports and parental appraisals moderate the effects of demands and stressors on family adaptation. This work also highlights the complex way in which the family functions and how the process of adaptation is active and includes the environment in which the family is situated and relationships with the wider community.

Further theoretical frameworks derived from the stress and coping literature have been developed which include the role that illness-related factors play in adaptation, and conceptualise the interrelationships of biomedical factors, psychosocial factors, and adaptation. The transactional stress and coping model is one such model (Thompson, Gil, Burbach, Keith, and Kinney, 1993), which has its roots in ecological-systems theory (Bronfrenbrenner, 1977) and conceptualises chronic illness as a stressor to which the individual and their family must adapt. The model includes biomedical indices and demographic parameters, but its focus is on individual and family psychosocial variables and how they mediate the illness-outcome relationship. It is hypothesised that the levels of stress and symptoms experienced by other family members impacts upon the psychological adjustment of the chronically ill child, and child adjustment is conceptualised as being interrelated with maternal adjustment. Mediation processes important to maternal adjustment include appraisals of stress, expectations of locus of control and efficacy, coping methods and family functioning. For the child, cognitive processes of expectations about self-esteem and health locus of control and methods of coping are seen as important. The model is explicitly

developmental and provides a framework for exploring the stability and change in adjustment and the interrelationship between maternal and child and family mediational processes over time (Thompson and Gustafson, 1996). A significant amount of the variance in psychosocial adaptation among children with sickle cell disease, cystic fibrosis, and spina bifida has been related to variables that are consistent with this model (Thompson, Gustafson, Bonner, and Ware, 1992; Thompson et al., 1993; Thomson, Gustafson, George, and Spock, 1994).

A similar model which also adopts a biopsychosocial approach to adaptation is the disability stress-coping model (Varni and Wallander, 1988; Wallander, Varni, Babani, Banis and Wilcox, 1989; Wallander and Varni, 1992). This model however, organises the factors proposed as contributing to adjustment to chronic illness in a risk-resistance framework. Adaptation is considered as a multi-dimensional construct, which includes mental health, social functioning, and physical health, and the major risk factor for developing psychosocial problems in children with chronic conditions is purported to be stress. Disease or condition parameters, functional limitations, and the psychosocial impact of the condition on the individual can be sources of stress. In addition to risk factors however, the model proposes three types of resistance factor, which come under the umbrella of coping resources and are conceptualised as being important to adaptation. *Intra-personal factors* include internal resources such as temperament, competence, and problem-solving ability. *Stress processing factors* include cognitive appraisal and coping behaviours. *Social-ecological factors*, the inclusion of which has been suggested as being this model's strength (Thompson and Gustafson, 1996), include family resources, such as the family environment, social support, and family members' adaptation (Wallander et al., 1989).

This conceptual model, illustrated in Figure 2-1 (p.27), was proposed to account for the differential psychosocial adjustment in chronically ill and disabled children by conceptualising adjustment in terms of risk and resistance factors, and in continued investigation the model has been extended to include other resources, such as social-ecological factors, that might serve as resistance factors in child adjustment (e.g. Wallander, Varni, Babani, Banis et al., 1988; Wallander et al., 1989; Wallander and Varni, 1989). The development of this model has therefore comprised a number of pieces of empirical work that make up an extensive research programme, which has



explored the adjustment of children affected by a number of conditions, including juvenile rheumatoid arthritis, spina bifida, and cerebral palsy. Thus, the authors have worked from the premise that there are commonalities in the psychosocial implications of different chronic disorders, plus they have proposed limited sets of risk and resistance factors rather than proposing specific factors. This has enabled the model to be widely heuristically applied. Indeed, the current body of research that has drawn on this model has identified a number of psychosocial variables that contribute to the variability in adaptation (a summary of which is given below) and it has been applied by numerous researchers to adaptation among children with a variety of chronic illnesses, including sickle cell disease (e.g. Brown, Doepke, and Kaslow, 1993), juvenile rheumatoid arthritis (e.g. Manuel, 2001), diabetes, and spina bifida (e.g. Wallander and Varni, 1992; Wallander et al., 1989).

Amongst individuals affected by MPS IH who have undergone bone marrow transplantation however, the matter of whether psychosocial resistance factors buffer the relationship between biomedical risk factors and adaptation has gone unexplored. It is therefore the objective of this research to examine the moderating effects of psychosocial factors on the relationship between biomedical risk factors and the psychosocial functioning of individuals affected by MPS IH post-BMT. It is also the objective of this research to apply the disability stress-coping model in its entirety to both parent and child adjustment. In the originating authors' investigations using this model, risk and resistance factors have been investigated piecemeal, and while they highlight the psychological morbidity of chronically ill children (Wallander and Varni, 1998) and illuminate the moderating effects of a number of resistance factors such as social support (Wallander and Varni, 1989) and psychological and utilitarian family resources (Wallander et al., 1989), no single study appears to have explored the relationships between variables in all of the risk and resistance factor sets proposed. This is also evident in numerous other pieces of research that have employed a risk-resistance framework. While they highlight the moderating and mediating effects of a number of intrapersonal, social-ecological, and stress processing variables on both parent and child stress, few appear to apply all parts of the model at the same time and look at both parent and child adjustment as being interlinked.

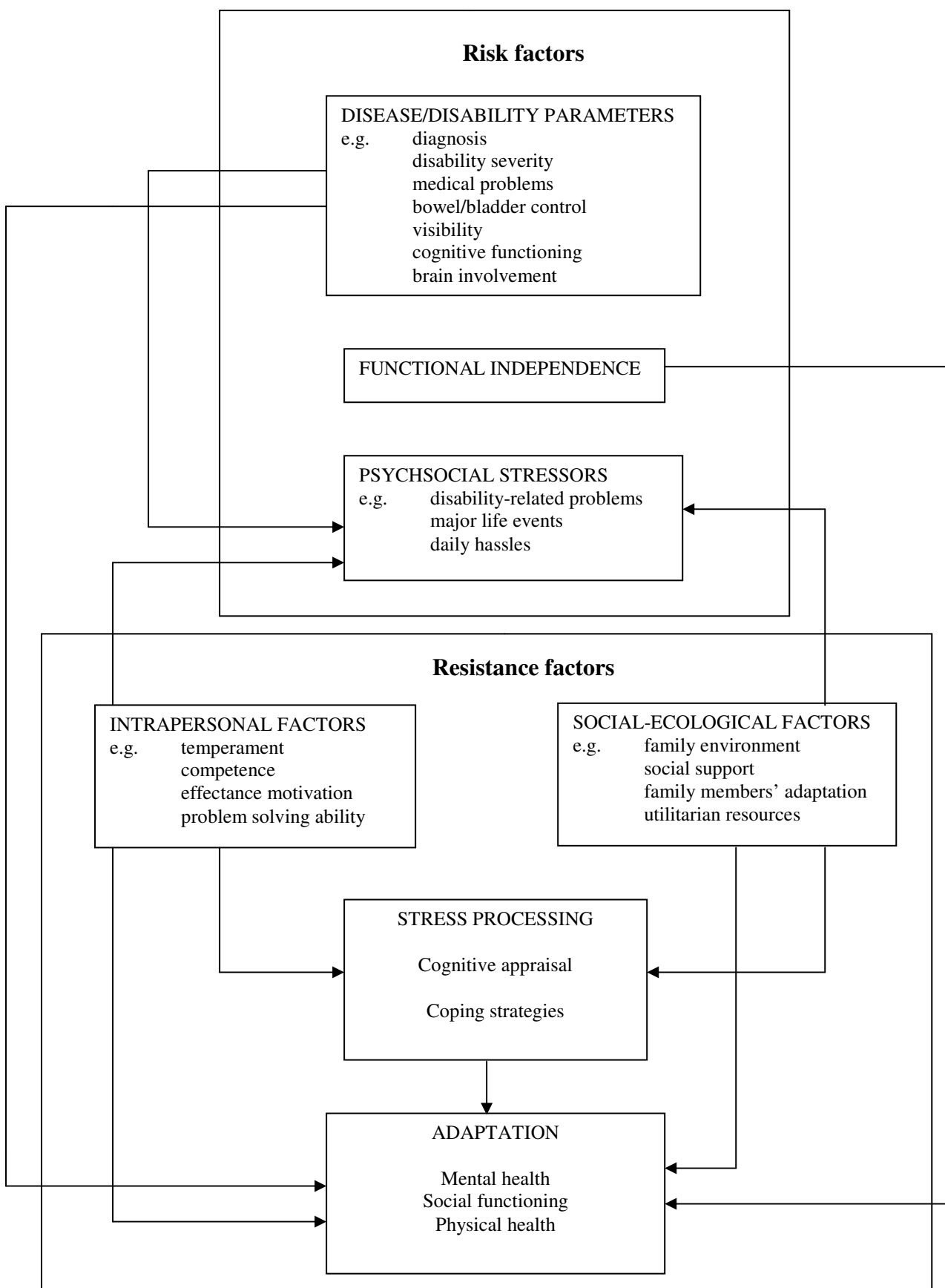
In terms of family adaptation to having a chronically ill or disabled child, previous research has explored a number of psychosocial resistance factors. Intra-personal resistance factors that have been identified amongst parents include mastery and control (e.g. Deatrick, Knafl, and Walsh, 1988; Gibson, 1988; Sigmon, Stanton, and Snyder, 1995), and normalisation and positive reframing (e.g. Eiser, 1990; Hill, 1994; Ray, 2002). Family functioning (e.g. Moos and Moos, 2002; Warfield et al., 1999), spousal support (e.g. Ray, 2002), intra- and extra-familial social support (e.g. Barakat and Linney, 1992; Oka and Ueda, 1998), and a variety of socioeconomic and family characteristic factors (e.g. Canning, Harris, and Kelleher, 1996; Wallander et al., 1989) are examples of social-ecological factors that have been explored in the field of childhood chronic illness. In terms of stress processing factors parental perceptions of the impact of the illness on the family (e.g. Ireys and Silver, 1996; Lustig, Ireys, Sills, and Walsh, 1996), sense of coherence (Olsson and Hwang, 2002), and perceptions of child vulnerability (e.g. Anthony, Gil, and Schanberg, 2003) have been widely studied and linked to the adjustment of parents of chronically ill children. In terms of risk factors, while disease severity has been found to have little impact on maternal and child adjustment (e.g. Canning et al., 1996; Wallander et al., 1989; Wallander and Varni, 1998), functional status, functional dependence, and everyday and illness-related stresses have shown stronger relationships to both individual and family psychosocial outcomes (e.g. Thompson et al., 1993; Wallander and Varni, 1992; Witt, Riley, and Coiro, 2003).

From the child's perspective, self-esteem (e.g. Appleton, Ellis, Minchom, Lawson et al., 1997; Thompson et al., 1994; Thompson et al., 1992) and self-concept (e.g. King, Shultz, Steel, Gilpin et al., 1993; Skar, 2003) are examples of intrapersonal factors that have been explored with the adaptation of individuals with chronic conditions such as sickle cell disease, spina bifida and cerebral palsy. The use of social support as a coping mechanism (Burlew, Telfair, Colangelo, and Wright, 2000), having a positive outlook (Livneh, Lott, and Antonak, 2004), and social relations (LaGreca and Bearman, 2000) are further protective and stress processing factors demonstrated to be associated with adaptation to chronic conditions in previous research. The most frequently studied social-ecological variables are family factors (Thompson et al., 1993). Previous research suggests that, among other factors, family support (Varni, Setoguchi, Rappaport and Talbot, 1991), childrearing behaviours (McAnarney, 1985;

Orr, Weller, Satterwhite and Pless , 1984; Thompson, Gustafson, Bonner, and Ware, 2002), parent distress (Thompson et al., 2002) maternal adjustment (Sawyer, Streiner, Antoniou, Toogood et al., 1998), family cohesion (Kliewer and Lewis, 1995), the parent-child relationship (Hurtig, Koepke, and Park, 1989), and organisation and control (Moos and Moos, 2002) may be important to our understanding of adaptation as they may serve as moderators and may affect adaptation in both direct and indirect ways.

With reference to the present study, as discussed the current body of research that has drawn on risk-resistance models of adaptation has identified a number of psychosocial variables that contribute to the variability in adaptation to chronic illness, and research associated with these models has made a significant contribution by doing so. It is therefore the objective of this research to examine the moderating effects of psychosocial factors on the relationship between biomedical risk factors and the psychosocial functioning of individuals affected by MPS IH post-BMT. It is hoped that this will elucidate factors that might moderate individual and family adaptation. To facilitate this, the exploration of previous research in the field of adaptation to childhood chronic illness will be explored in this thesis from a risk-resistance perspective and set out in line with the disability stress-coping model (Wallander et al., 1989; Wallander and Varni, 1992). This model provides a useful framework from which to explore a childhood disorder for the first time. It allows for the potential disease- and disability-related risk factors to be identified, and then for the resistance factors to be systematically explored in terms of intra-personal, social-ecological, and stress processing factors. As previously mentioned, it is the inclusion of additional social-ecological factors in this model that give it its strength over other models of adaptation to chronic illness (Thompson and Gustafson, 1996), and are felt to be of critical importance to both parent and child adjustment.

**Figure 2-1. The Disability Stress-Coping Model**



Reproduced from Wallander, Varni, Babani, Banis, and Wilcox, 1989

## **2.1. Living with Chronic Illness and Disability: The Impact on the Family**

A rich history of research into the concept of childhood resilience has highlighted the family system as integral. When children face risk situations a number of parenting and family systems factors have been shown to serve as protective factors that enhance resilience. These include consistent discipline and optimism regarding the child's future (Wyman, Cowen, Work, and Parker, 1991), maternal social support (Conrad and Hammen, 1993), stable, consistent, and nurturing family environments (Wyman, Cohen, Work, Raoof et al., 1992), and supportive relationships (Werner and Smith, 1982). As well as the family environment, individual characteristics of the child and factors related to the wider social context also serve as protective factors and interact with one another (Cohler, 1987). Furthermore, the balance between the stressful life events that intensify children's vulnerability and the protective factors that heighten resilience shift across time as the child and other family members grow and develop over the life cycle. The balance also varies according to the child's gender and the social and cultural context in which the child matures (Werner and Smith, 1992).

Research exploring childhood chronic illness has produced findings that are consistent with that which has explored resiliency in childhood more widely. The psychosocial adjustment of children affected by chronic illness or disability has been more strongly related to individual, parent, and family related factors than to illness or disability parameters alone (Lavigne and Faier-Routman, 1993). This will be explored in more depth in the next section. In the meantime, as family functioning and other social ecological factors have been strongly linked to child adaptation to chronic illness, the importance of exploring the factors that increase *family* resilience in the face of stressful life events is supported. It is of great import to this research that an understanding of the conditions that lead families into a crisis, and an understanding of the factors that shape parents' ability to cope, is sought. For the purpose of clarity, events that are considered to be crisis-producing are defined as events that:

‘disrupt the family system and that precipitate changes in....the family’s patterns of functioning, thus placing the family system at risk for continued decline in functioning leading to dysfunction’ (McCubbin, McCubbin, Thompson, Han et al., 1997:4).

Of the three classes of stressors that affect families and require management (daily hassles, developmental transitions, and cataclysmic events) chronic illness of a family member is recognised as a cataclysmic event (Kiser, Ostojja, and Pruitt, 1998) and the diagnosis of chronic illness in childhood constitutes a major family crisis (Eiser, 1990).

Although parents are at risk of adjustment difficulties when they have a child with chronic illness, research exploring the impact it has on the family suggests that pathological functioning among this group is not usually found (Hanson, 2001; Wallander and Varni, 1998). Risk-resistance models of stress and coping provide a useful framework for understanding the way in which families can be overwhelmed and severely challenged in the face of adversity, but have the ability to recover and adapt to the situation subject to a number of intrapersonal, stress processing, and social-ecological factors, which buffer the impact the illness has on the family (Wallander et al., 1989; Wallander and Varni, 1992). It is also useful to view coping as a non-linear process, which involves a series of ‘ups and downs’ (Spinetta, 1981) and unexpected challenges (Wortman and Silver, 1987), which require the family to go through repeated processes of recovery and adaptation. For more than two decades research on families under stress has explored a number of variables that have been conceptualised as protective factors that operate within family systems when crisis situations and risk factors present themselves. A number of these observations will be explored in the following section in relation to childhood chronic illness and disability using the disability stress-coping model’s risk-resistance framework (Wallander et al., 1989; Wallander and Varni, 1992). Before exploring resistance factors however, a number of risk factors will first be examined. A number of risk factors for adjustment problems among parents of chronically ill children have been identified, including disease parameters, functional care strain, and psychosocial stress (Wallander and Varni, 1998).

### **2.1.1. Risk Factors**

For most families ‘diagnosis of chronic illness comes as a shock and gives rise to issues of loss (of a healthy child and his or her future), disbelief, denial, anger, and in some cases guilt (especially if the condition is genetically transmitted)’ (Kiser et al., 1998:95). Additional reactions include sadness, despair, depression, helplessness, frustration, confusion, and conflict (Cohen, 1993; Jerret, 1994; Eakes, 1995). Families can also experience a sense of ambiguous loss and disenfranchised grief, particularly if a family member has been diagnosed with a genetic condition but is asymptomatic (Sobel and Cowen, 2003). Many families also find the demands of caring for a child that is chronically ill to be extremely taxing, physically, financially, and emotionally (Gallo, 1991; Swallow and Thompson, 1992; Ray and Ritchie, 1993; Williams, Lorenzo, and Borja, 1993; Stewart, Richie, Mcgrath, Thompson, and Bruce, 1994; Gibson, 1995), and parents of chronically ill children often experience higher levels of psychological stress (e.g. depression and anxiety) than parents of healthy children (Ievers and Drotar, 1996). The stressors experienced by parents of children who are chronically ill are multiple and ongoing, and vary over time. From a parental experiential perspective they have been categorised as happening at the time of diagnosis and throughout developmental transitions, and to be related to the ongoing health care needs of the child, including when the child experiences illness exacerbations and hospitalisations (Melnik, Feinstein, Moldenhouer, and Small, 2001).

#### **2.1.1.1. Psychosocial Risk Factors**

Having a child diagnosed with a chronic illness constitutes a major family crisis (Eiser, 1990) and as well as a number of emotional responses as described above parents can also experience decreased self-worth and a lack of confidence (Stevens, 1994), and can experience a mourning reaction (McCollum and Gibson, 1970). Since chronic illness may negatively affect the physical, cognitive, and adaptive functioning of the child, and/or their emotional health, successful transition through developmental milestones may be challenged. This can be an extremely painful experience for parents as they watch their child struggle to accomplish developmentally appropriate tasks, especially when differences and delays between

their children and healthy peers become apparent (Melnyk et al., 2001; Thompson and Gustafson, 1996). This can give rise to recurrent episodes of chronic sorrow (Clubb, 1991) and can be 'trigger points' for uncertainty (Cohen, 1995). Chronic sorrow was originally described in the literature as a coping mechanism that allows for periodic grieving (Olshansky, 1962). Rather than a continuous process, it is thought to be a recurrent phenomenon (Winkler, 1981), which forms part of the adaptation process. It is suggested that parents go through a series of stages of adaptation to their child's illness over time. For example, impact, denial, grief, focusing attention, and closure. However, they may re-experience peaks in the grieving process during developmental transitions and high-risk periods (Clubb, 1991).

Depending on the severity of the condition and whether cognitive function is impaired, high risk periods might include some or all of the major developmental transitions such as walking (12-15 months), talking (24-30 months), the age of entry to school, the onset of adolescence, and the transition to adulthood. Other high risk periods relate to disease-specific events, such as the time of diagnosis and occasions when professional interventions are needed (Winkler, 1981). Uncertainty, which has been related to family distress (Cohen, 1993, 1993a), can be further exacerbated when disease symptoms are intermittently unpredictable (Dodgson, Garwick, Blozis, Patterson, Bennett, and Blum, 2000). Normal developmental transitions such as starting school are more stressful for mothers of chronically ill children than mothers of healthy children (Thompson and Gustafson, 1996). The transition to school may illuminate for parents their child's difference to their peers in terms of appearance, social skills, and cognitive and physical functioning. As well as seeing their child challenged by age-appropriate activities, parents may also have to watch their child struggle socially and experience difficulties establishing friendships (Trachtenberg and Batshaw, 1997). When the child enters the school system, parents also relinquish the day-to-day health care management of their child's condition to school personnel. Giving up control of this element of their child's care is an added source of stress and concern for parents, not least when 'many teachers and school personnel have very little knowledge about childhood chronic illnesses' (Melnyk et al., 2001:3). Teachers' lack of understanding about a condition can also lead them to have expectations of the child that are either too high or too low, which can be distressing for parents (Brewer, Eatough, Smith, Stanley et al., 2008). Adolescence is also a stressful time for families



affected by childhood chronic illness. Allowing the chronically ill adolescent to have increased autonomy and independence, while maintaining adequate supervision, support, and nurturing care is a major challenge for parents (McAnarney, 1985).

While historically much research exploring the impact that childhood chronic illness has on the family has focused on the psychological adjustment of the mother (see Bailey, Blasco and Simeonson, 1992), more recent research has shown how the experience can impact on mothers and fathers in different ways. When stressful life events affect family and friends, women have been found to be more prone to psychological distress than men (e.g. Aneshensel, 1992; Gadzella, Ginther, Tomalca and Bryant, 1991; Thoits, 1995), and mothers of children with chronic illness and disability have been found to experience higher levels of stress and depression than their spouses (e.g. Beckman, 1991; Timko, Stovel and Moos, 1992; Sloper and Turner, 1993; Pelchat, Ricard, Bouchard, Perreault et al., 1999a; Pelchat, Bisson, Ricard, Perreault, et al., 1999b). It is purported however, that reactions to stressful life events are more likely to be related to gender roles and the stressful events that such roles demand, than simply to the sex of the people involved (Banyard and Graham-Berman, 1993; Gray, 2003). For example, it is commonly reported that women are more likely than men to take most responsibility for the care of sick or disabled family members (Anderson and Elfert, 1989; Guberman, Mahue and Maille, 1992; Parks and Pilisuk, 1991) and to simultaneously take care of the upkeep of the home whether or not both parents work (Pelchat, Lefebvre and Perreault, 2003). Research has also found mothers to deal with illness-related stressors such as medication regimens and special transportation needs (Thompson et al., 1993) and to usually liaise between the family and medical practitioners (Goldner, 1985). Furthermore, mothers also deal with the routine stresses of everyday life, which have themselves been directly related to the adjustment of mothers of chronically ill children (Thompson et al., 1993b). It is suggested then that 'the additive effects of illness-related and ordinary hassles may cause increased difficulties for mothers' (p238) when they have a chronically ill child, and psychosocial stress resulting from chronic illness is suggested to be a significant source of adjustment problems in mothers of chronically ill children (Manuel, 2001).

Illness and disability in the family has also been shown to hold different significance for men and women. It is asserted that, while the day-to-day demands of caring for a child with disabilities make adapting more difficult for mothers, the difficulties fathers have in adapting relate to the disability itself (Krauss, 1993; Keller and Honig, 2004) and to the impact the illness has on the family (Pelchat et al., 2003). Fathers' stress has been related to their child's disposition (Krauss, 1993); their child's social acceptability (Saloviita, Italinna and Leinonen, 2003; Keller and Honig, 2004); their relationship with their child (Krauss, 1993; Cohen, 1999); the financial burden of meeting their child's needs (Brown and Barbarin, 1996; Cohen, 1999); and the time impact on the marital relationship (Heaman, 1995). They are also reported to recognise their spouse's over-involvement with the child and are to have concerns about the marital relationship and about other children in the family, seeking to encourage their spouse to spend time with them and other family members (Pelchat et al., 2003). The differences in parents' concerns is purported to be due to mothers' and fathers' parenting roles and the different ways in which they interact with their children. Mothers are ostensibly more emotionally involved with their child than fathers and their concerns have been found to centre around their abilities as a parent and the care of their child. Fathers on the other hand are said to be involved on a more cognitive level (Pelchat et al., 2003). Overall, fathers do not interact with their children as frequently as mothers, and their role is not as focussed on care-giving as is a mother's (Fagot, 1995; Harris, Furstenberg and Marmer, 1998). Fathers tend to engage in more physical activities with their children (Collins and Russell, 1991; Forehand and Nousiainen, 1993; Parke 1996); and encourage traits like independence and competitiveness more so than mothers (Cabrera, Tamis-LeMonda, Bradley, Hofferth et al., 2000). In some instances, when they have a child that is chronically ill or disabled, fathers can experience difficulties recognising and adjusting to their role (Stork, 1995), and in some cases can distance themselves from being 'fatherly' (Neyrand, 2000). Since having a chronically ill child can differentially impact on mothers and fathers, both emotionally and physically, it is important to consider both parents' experiences when evaluating family adaptation. The father- as well as the mother-child relationship has been demonstrated as being important to child adjustment (Rohner and Veneziano, 2001), and efforts to understand both parents' needs are therefore key to helping minimise stressful and distressing experiences associated with care-giving.

### **2.1.1.2. Illness-Related Risk Factors**

In terms of illness-related risk factors, research has investigated the relationship between specific aetiologies or aspects of chronic childhood conditions and parental adjustment. There has been little consensus however. For example, chronic illnesses which have central nervous system involvement and cognitive impairment and which are severely handicapping have been related to more negative outcomes for the family (Breslau, Staruch, and Mortimer, 1982). It is suggested that this is due to such conditions being more functionally disabling than others (e.g. Kovacs, Finkelstein, Feinberg, Crouse-Novak et al., 1985), and the fact that they can present with behavioural difficulties which can be stressful to manage (Hodapp, Wijma, and Masino, 1997). Studies that have explored specific genetic disorders indicate that rates of family stress are related to the condition by which their child is affected. For example, families with a child affected by Downs Syndrome have been reported as showing lower levels of stress than families of children with other disorders such as autism (Kasari and Sigman, 1997). One reason offered for the comparatively low stress levels shown by these families is parental support and knowledge of the condition (Hodapp, 1995). In support of this, research has demonstrated how parent illness-related stress is exacerbated by lack of knowledge about a condition (e.g. Ievers and Drotar, 1996; Brewer et al., 2008). The age of the child has also been suggested as a factor that contributes to parenting stress. The parenting of young children with developmental disabilities is purported to be more stressful than the parenting of older children with similar disabilities (Baxter, Cummins, and Polak, 2000; Beckman, 1991). This is not a consistent finding however (Flynt and Wood, 1989) as ongoing care is also highlighted as a significant source of stress for parents (e.g. Ievers and Drotar, 1996), and the duration of a child's illness has been related to maternal psychosocial functioning (Timko et al., 1992).

Among parents of children affected by chronic conditions such as cystic fibrosis and type I diabetes a commonly cited source of stress is commitment to the day-to-day treatment regimens associated with complications of the condition (e.g. Ievers and Drotar, 1996; Sullivan-Bolyai, Deatrick, Gruppuso, Tamborlane, and Grey, 2003). For many chronic conditions, parents are involved in the regular provision of complex

therapies to their children, including intravenous drug therapies, physiotherapy, dietary management, blood glucose monitoring, and insulin administration (e.g. Moore, 1988; Ellis, 1989; Stephenson, 1989; Boland and Grey, 2000; Canam, 1993), and families in effect become part of a multi-disciplinary team working collaboratively to provide care for their child (Coyne, 1997). Parents of chronically ill children report having concerns about their competence in providing adequate care, particularly in the months following diagnosis (e.g. Hatton, Canam, Thorne, and Hughes, 1995; Sullivan-Bolyai et al., 2003). Finding a balance between the management of the child's condition, feelings of fear about the child's welfare, and meeting the developmental needs of the child is also a source of stress for parents (e.g. Sullivan-Bolyai et al., 2003). Furthermore, the trajectory of many chronic illnesses can be interrupted by exacerbations and deterioration in the child's functioning, which put a strain on parents' coping resources. This results in frequent hospitalisations for the child, increased service use, and disruption to the family lifestyle and normal routines (Melnik et al., 2001). These episodes are also fraught with emotions as parents can feel a sense of powerlessness and lack of control (Faulkner, 1996), and a deep sense of uncertainty about their child's future well-being (Simon and Smith, 1992).

While illness-related stress has consistently been found to be a principal source of adjustment problems in mothers of chronically ill children (Thompson and Gustafson, 1996), the general consensus regarding illness severity is that it is not related to psychological outcomes for mothers (e.g. Wallander, Varni, Babani, Banis, DeHaan, et al., 1989; Thompson et al., 1992; Canning et al., 1996). The child's functional status however, is suggested as having a much larger impact on maternal psychological functioning (e.g. Canning et al., 1996; Timko et al., 1992; Wallander and Varni, 1992), and functional dependence has been identified as a major cause of care strain amongst parents of chronically ill or disabled children (Pless and Pinkerton, 1975; Wallander et al., 1989; Timko et al., 1992; Wallander and Varni, 1992; Wallander and Varni, 1998). Distinctions have been made between illness and functional severity (Stein, Gortmaker, Perris, Perrin, Pless, and Waler, 1987). Illness severity refers to disease-specific physiological symptoms (Lustig et al., 1996). Illness severity for MPS IH post-BMT may include severe joint pain, back pain, stiffness, and limitation in motion. It may also include GvHD-related symptoms such as lung

disease. Functional severity reflects the impact the disorder has on physical, adaptive, and social functioning (Lustig et al., 1996), including impairments in activities of daily living and limitations in intellectual and emotional functioning (Billings, Moos, Miller, and Gottlieb, 1987; Daniels, Moos, Billings, and Miller, 1987; Stein et al., 1987). Children affected by MPS IH post-BMT experience significant and progressive limitation in physical functioning and often require assistance with activities of daily living. They have extremely poor eyesight and experience some degree of cognitive impairment, which can limit social and adaptive functioning. As previously discussed, the care of the child often falls to the mother (e.g. Anderson and Elfert, 1989; Faulkner, 1996; Guberman et al., 1992; Parks and Pilisuk, 1991), and balancing the competing demands from the child's chronic illness and other responsibilities of family life is a continual source of stress for them (Barlow, Harrison, and Shaw, 1998; Atkin and Ahman, 2000). Mothers of chronically ill children are therefore at an increased risk of anxiety, depression, role-strain, exacerbation of existing relationship problems, disorganisation of family routines, loneliness, and isolation (Venters, 1981; Wallander et al., 1990; Havermans and Eiser, 1991; Patterson et al., 1992; Gibson, 1995; Ievers and Drotar, 1996). They have also been found to experience greater parent distress when they feel unable to pursue personal interests and goals as a result of restrictive parenting responsibilities (Breslau et al., 1982).

Adjustment to chronic illness within the family also involves the development of a relationship between the family and the health care system, the quality of which may further affect one's ability to adjust to and cope with the condition (Heijmans, Foets, Rijken, Schreurs et al., 2001). Parents often describe difficulties communicating with health care providers at this early stage (Knafl, Ayres, Gallo, Zoeller, and Breitmayer, 1995), and unhelpful support from professionals has been highlighted as a source of stress for parents both at diagnosis and throughout the course of the condition (e.g. Brewer, Smith, Eatough, Stanley et al., 2007). The experience of being in a high-technology hospital environment has also been identified as an additional stressor for parents of children newly diagnosed with a chronic condition (Hughes and McCollum, 1994). Furthermore, ongoing care can place a financial burden on the family, which can be a major source of stress. Families often have to make adjustments to their lifestyle and modifications to their homes, which presents the

family with significant financial strains (Samuelson, Foltz and Foxall, 1992). Additional travel costs add to this burden. While government grants are available, families often compete for charitable support, and in some countries have to pay for medical care as insurance companies often do not meet the costs (Melnik et al., 2001).

The majority of research in this area focuses on specific conditions (Wallander and Varni, 1998) and there is some debate about whether this diagnosis-specific approach is more appropriate than one that looks at commonalities across conditions when examining the impact chronic illness has on families (Eiser, 1990, 1993). It is argued that a non-categorical approach 'can impede our in-depth understanding of the condition's impact on the family and the development of more specialist services' (Brewer et al., 2008:6). Conversely, it is believed that the psychological impact of chronic illness on children and families is the manifestation of the processes common to all chronic illness and may not be particular to the medical condition involved (Stein and Jessop, 1982a). This position is taken by Wallander and colleagues in their disability stress-coping model. This notwithstanding, conditions that are genetically transmitted are suggested as having particular implications for the family psychosocially and for family systems (Williamson, 1999). For example, research has demonstrated how the genetic nature of a child's condition can give rise to feelings of guilt and self-blame in parents (e.g. Brewer et al., 2008). When conditions are rare there is also a lack of knowledge and understanding from family members, professionals encountered by parents, and from society at large. As previously mentioned teachers can have unrealistic expectations of children (Brewer et al., 2008), extended family members can feel uncomfortable with disability and withdraw contact (Ray, 2002; Pelchat et al., 2003), inappropriate advice or misinformation can be offered (Suls, 1982), and inappropriate attributions made for child behaviour which imply that the parent is to blame (Brewer et al., 2007). Lack of information or understanding about a condition is a significant source of stress to parents of children with chronic illness and can lead to feelings of isolation (e.g. Brewer et al., 2008; Ievers and Drotar, 1996). The parent therefore becomes the 'expert', having to inform teachers, support services, and family and friends alike about the child's condition, its cause and manifestations, and about the needs of the child.

There is a growing body of work that demonstrates that the majority of parents adjust well and successfully handle the demands placed on them as a result of having a chronically ill child (e.g. Kellerman, Zelter, Ellenberg, Dash et al., 1980; Drotar, Doershuk, Stern, Boat et al., 1981; McCubbin, 1984; Sinnema, 1984; Cowen, Corey, Keenan, Simmons et al., 1985; Knafl and Deatrick, 1986; Walker, Ford, and Donald, 1987; Gibson, 1986; Gibson, 1988; Cappelli, McGrath, MacDonald, Katsanis et al., 1989; Sawyer, 1992; Davies, 1993). That said, the enormous burden that caring for a chronically ill child can put on the family must not be underestimated. As previously mentioned, when a child is chronically ill, the bulk of the care often falls to the mother, and this can lead to psychosocial adjustment problems as discussed. Equally, when a child is seriously ill, it can be difficult for a parent to separate themselves from the caring role and from their child's pain and suffering and they can often be engulfed by the situation (Twigg and Atkin, 1994). However, a number of resistance or protective factors have been found to buffer the effects that illness parameters, functional care-strain, and psychosocial factors have on the psychological adjustment of parents of chronically ill children. These factors have been described by the disability stress-coping model as intrapersonal, social-ecological, or stress processing (Wallander and Varni, 1992), which will be explored in the following section.

### **2.1.2. Resistance Factors**

Little research has explored the effects that intrapersonal factors have on parents of chronically ill children. This may be due to the belief that these factors are stable and not open to intervention (Thomson and Gustafson, 1996). However, the research that has been carried out suggests that they are important to well-being (e.g. Silver, Bauman, and Ireys, 1995) and the continued examination of the effects intrapersonal factors have on the relationship between stress and adaptation is important. Social-ecological factors on the other hand have been widely researched, and family factors have been strongly related to both maternal and paternal adjustment (Barakat and Linney, 1992; Heaman, 1995; Canning et al., 1996; Pelchat et al., 2003). The importance of social support to maternal adjustment is also well documented, and a number of studies have reported the significance of both intra- and extra-familial social support to mothers' adaptation. Mothers' reports of increased family

supportiveness and larger support networks have been positively related to the adjustment of mothers of children affected by different illnesses and to that of their children (Barakat and Linney, 1992), and to mothers' mental and social functioning (Wallander, Varni, Babani, DeHaan et al., 1989). Other family resources, such as socioeconomic status and mother's education and age, have also been found to effect maternal stress (McCormick, Athreya, and Stemmler, 1985; Wallander et al., 1989; Canning et al., 1996).

#### **2.1.2.1. Intrapersonal Resistance Factors**

Internal resources such as self-esteem and self-mastery, which is defined as a general sense of personal control over life events (Pearlin and Schooler, 1978) have been positively associated with mothers' psychological adjustment to having a sick child. For example, mothers of special care newborns have been found to experience lower levels of depression and fewer major stress reactions when they feel a greater sense of personal control over their child's recovery (Affleck, Tennen, Allen, and Gershman, 1986). Mothers of well, acutely ill, and chronically ill children have also been reported to experience less psychological distress when they are high in self-mastery (Hobfoll and Lerman, 1988), and amongst mothers of children affected by a variety of chronic conditions less psychological distress has been associated with more positive views of self-worth and a greater sense of personal control over life events (Silver et al., 1995). Internal coping resources have also been positively related to family satisfaction (Snowdon, Cameron and Dunham, 1994) and self-esteem in particular has been associated with marital satisfaction and marital adaptation to having a chronically ill child. Mothers with a high self-esteem have been found to be able to appropriately differentiate between the needs of their child and those of their spousal relationship, and to sustain acceptable levels of marital satisfaction (Florian and Findler, 2001).

Other intrapersonal factors that have been found to protect individuals from the negative effects of stress include self-efficacy, self-confidence, and perceived control, and the personality disposition underlying these related variables is seen as particularly relevant to coping. Perceived self-efficacy pertains to the judgments one makes regarding their effectiveness in managing stressful or unpredictable situations,



and it is believed to predict a wide range of adaptive behaviour including coping responses, persistence, and achievement motivation (Bandura, 1982). As previously discussed, the day-to-day management of a childhood condition and the balancing of the caring role with other personal and family roles are inordinately stressful for parents. Active coping strategies such as the incorporation of illness management tasks into a schedule have been found to increase feelings of control (e.g. Deatrick et al., 1988; Gibson, 1988; Knafl, Breitmeyer, Gallo, and Zoeller, 1996). For many parents of chronically ill children, knowing they have the ability to manage the condition on a day-to-day basis has been reported as being more beneficial to them than seeking understanding of and explanations for the condition (Atkin and Ahmad, 2000). This not only helps families to achieving mastery, which ‘aims at controlling the manifestations of [disease] symptoms’ (Atkin and Ahmad, 2000:59), it also helps to normalise the experiences associated with having a chronically ill child as illness-related routines become part of the ‘normal’ family environment (Krulik, 1980). The organisation of care and other such normalising tactics have been found to help family adaptation (Krulik, 1980). They are further purported to help reduce the impact the illness has on the child and to promote the child’s integration into mainstream society by supporting their development and competence (Holaday, 1984). Furthermore, mothers who are more confident about their problem-solving abilities have been found to be more likely to opt for more active coping strategies and consequently to have more positive feelings toward parenting (Noojin and Wallander, 1997), and perceived self-efficacy has been related to parents’ reports of adaptive family functioning (Kazak, McClure Alderfer, Hwang et al., 2004).

The development of routines and coming to terms with the realities of what is involved in caring for a chronically ill child can take some considerable time however, often extending to several months or even years (Venters, 1981; Yarcheski, 1988; Whyte, 1994). Taking things step by step however, offers parents some protection against ‘engulfment’ (Twigg and Atkin, 1994) or being overwhelmed by thoughts about an uncertain future and a lifetime of burden (Atkin and Ahmad, 2000). It is seen as a practical response, which allows families to deal with problems as they arise and focus on their ability to manage, thus offering mastery of the condition (Hill, 1994; Jerrett, 1994; Atkin and Ahmad, 2000). It also allows parents to establish routines and mastery of treatment regimens over time (e.g. Deatrick et al., 1988;

Sullivan-Bolyai et al., 2003) and to re-evaluate illness management behaviours as the child's developmental needs change (Sullivan-Bolyai et al., 2003). Often parents use the expression '*living one day at a time*' (Ray, 2002), which illustrates the ongoing process of adaptation and the way in which illness representations are constantly modified as families experience living with the condition and appraise coping strategies (Leventhal et al., 1997). A number of strategies are used to enable families to remain positive about their situation and gain some control over the condition, and optimistic interpretation of the situation and focusing on the present has been found to enable parents to be hopeful about the future (Venters, 1981). Family adaptation to childhood chronic illness is constantly evolving and subject to fluctuations. It is mediated by a number of intrapersonal resistance factors as discussed, but it is also subject to further moderation by a number of social-ecological factors including family functioning, family and social support systems, and other family resources.

#### **2.1.2.2. Social-Ecological Resistance Factors**

##### **2.1.2.2.1. Family Functioning**

It is reported that some marriages become weakened by the stress of chronic illness or disability in the family, 'as they become vulnerable to tension, conflict, blame, and resentment' (Kiser et al., 1998:94). Additionally, other relationships in the family may become affected, including that of the parents and other children in the family (Quittner, 1994). However, while relationships may be damaged in some families, for others the experience can be a positive and rewarding one and in some instances marriages become strengthened by the experience (Candy, Davies, and Ross, 2001). Again, the ways in which family members cope with such crises depend upon a number of intrapersonal, social-ecological, and stress-processing factors (Wallander et al., 1989), and the functioning of the family as a unit is crucial to adjustment (Warfield et al., 1999). A number of studies have explored a variety of elements of family functioning, and found relationships between them and parental coping and adjustment to having a chronically ill child. For example, although an increase in structure is often found in families of children with disabilities it depicts organisation orientation and is seen as an effective coping strategy (Moos and Moos, 2002).

Families who exercise a stable routine are often found to be high in organisation and control, low in conflict, and high in cohesion orientation (Fiese and Kline, 1993). Families that are orientated towards recreational pursuits are also reported as having more friends and acquaintances than families that are less inclined to take part in such pursuits (Moos and Moos, 2002).

Family cohesion and the quality of the marital relationship are two family functioning variables that have been found to hold particular relevance to family adaptation to childhood chronic illness (Wallander and Varni, 1998). Family cohesion has been associated with reports of increased supportive behaviours from family members (Sandler and Berrera, 1984), more parental care and less parental overprotection (Sarason, Shearin, Pierce, and Sarason, 1987), and increased parental reliance on problem-solving coping strategies (McCubbin, McCubbin, Patterson, Cauble et al., 1983). It is further cited as being instrumental in families' acceptance of their child's disabilities and in their coping competency (Bristol, 1984), and associated with greater social support (Sarason et al., 1987). Moreover, mothers of chronically ill children are at particular risk of parenting and adjustment difficulties when the family environment is not particularly cohesive (Warfield et al., 1999). In terms of the marital relationship, spousal support is cited as being more predictive of maternal adjustment to childhood chronic illness than illness parameters (McCubbin and Patterson, 1983), and it has been seen to protect mothers from stress, especially when fathers highly value child-rearing activities (Nagy and Ungerer, 1990). Spousal support is therefore reported as being the most important type of support for mothers (Ray, 2002) and highlighted as an important factor in family adaptation for both mothers and fathers (Goldberg, Marcovitch, MacGregor and Lojkasek, 1986; Grant and Whittell, 2000).

However, as previously discussed, mothers and fathers are differentially impacted upon by childhood chronic illness and respond differently in line with their parenting and social roles (Gray, 2003). They can also have different ideas about how the family works, the effectiveness of their communication and coping styles, and can have different concerns regarding the care of their chronically ill child (Crowley and Taylor, 1994; Pelchat et al., 2003). This can have implications for family functioning and intra-familial supportive behaviours. For example, mothers are particularly

concerned with the care of their child, their well-being, and their role as parent in the child's adaptation (Pelchat et al., 2003). They also take most responsibility for the day-to-day responsibilities associated with the care the child as well as everyday family responsibilities (e.g. Parks and Pilisuk, 1991; Pelchat et al., 2003; Thompson et al., 1993). Consequently, the lack of family support has been reported as a strong predictor of stress amongst mothers of chronically ill children (Margalit, Leyser, Avraham, Lewy-Osin et al., 1988; Atkin and Ahmad, 2000). Conversely, fathers' adjustment can be particularly affected by the impact the child's condition has on the family (Pelchat et al., 2003), its time impact on the marital relationship (Heaman, 1995), and a strong predictor of distress amongst fathers is purported to be the lack of personal growth orientation within the family environment (Margalit, Leyser, Avraham, Lewy-Osin et al., 1989). Fathers are also reported as being concerned with their spouse's over-involvement with the chronically ill child (Pelchat et al., 2003), which can exclude him from the caring role and minimise his involvement (Lillie, 1993).

Men and women are also suggested to use different coping strategies and to express themselves in different ways. Mastery and control have been espoused as useful coping resources, which are used more by men than women (Sigmon et al., 1995). Men are also known to use a stoical and inexpressive style of coping with stressful events, while women use a more emotional and expressive style and are more likely to seek support from family and friends (Thoits, 1995). These are examples of 'emotion-focused' and 'problem-focused' coping (Borden and Berlin, 1990; Kvam and Lyons, 1991; Lazarus, 1993, 1996; Sigmon et al., 1995). The ways in which parents express themselves however, has been reported as being perceived differently by mothers and fathers. Mothers have been reported as finding their partner's reluctance to express emotion as problematic, while fathers have reported feeling that they do discuss issues relating to their child with their spouse, and that they do not see their communication methods as problematic (Pelchat et al., 2003). It is posited that 'these contradictions make it evident that mothers and fathers have different standards for evaluating their own communication and that of others, and have different perceptions of what communication and dialogue are' (p239). Indeed, parental communication difficulties have been associated with parents' ability to cope with their child's chronic illness (Whyte, 1992).

Failure to understand how mothers and fathers differentially experience and cope with having a chronically ill child is therefore important to spouses on a number of levels. Not least because these factors determine the kind of support parents give to and receive from one another. It determines whether or not support received is 'in tune' with expectations, which determines whether or not it is perceived as helpful (Bristol, Gallagher, and Schopler, 1988). Family functioning and the quality of the marital relationship is vital to parent adjustment to having a chronically ill child, and being able to share the burden is a coping strategy that has been associated with optimum family functioning (Venters, 1981; McCubbin, 1984). By establishing a support system within the family that shares the burden of the illness the family's vulnerability to stress can be reduced (Holaday, 1984). It is purported however, that for family support to be effective it is necessary for decision-making to be shared, parents to adopt a variety of parental roles, and each member of the family to be recognised as important contributors to the family unit (Holaday, 1984). Strong, close relationships have been found to protect against the most negative effects of stress (e.g. Hobfoll and Walfisch, 1984), and perceived as well as actual support is a protective factor (Leatham and Duck, 1990; Sarason et al., 1997). Since it has been suggested that the quality of the marital relationship is related to the well-being of the parents and to the quality of care that is given to the child (Belsky, 1981) and family functioning an important contributor to child adjustment (Drotar, 1997), the dynamic interplay between support offered, expectations of and satisfaction with support received plays an important role in family adjustment to childhood chronic illness.

#### **2.1.2.2.2. Family Resources**

A number of family resources are also suggested as being related to parental adjustment to having a chronically ill child. Characteristics of the family such as income, mother's education and age, and duration of marriage have been related to maternal stress (Wallander et al., 1998), and in turn utilitarian family resources have been related to child psychosocial outcomes (Wallander et al., 1989). Low family income and maternal education have, in particular, been related to maternal distress (McCormick et al., 1985; Canning et al., 1996). While social status has been highlighted as an important determinant of psychological well-being (Kessler, Price,

and Wortman, 1985), parents of chronically ill children are ostensibly more vulnerable to fluctuations in their financial resources regardless of social status. The illness itself and the non-linear nature of many chronic conditions are likely to place demand, sometimes unexpected, on parents' finances (Canning et al., 1996). With reference to maternal education, its relationship with maternal adjustment is likely to be an indirect one. It is suggested that more highly educated mothers are better able to access social resources and information, have more knowledge of resources, and to be more organised. It is suggested that such resources enable mothers to more adequately manage 'tangible everyday nuisances' before they impact on them psychologically (Manuel, 2001). Lower levels of maternal education on the other hand have been related to perceptions of child vulnerability (Anthony et al., 2003), which has more direct implications for maternal adjustment (e.g. Cohen, 1997). Perceptions of child vulnerability have been associated with poorer adjustment for both mothers and children (e.g. Stewart and Mishel, 2000; Anthony et al., 2003), which will be discussed in the next section. Maternal knowledge about child development (Thompson et al., 2002) and cognitive growth fostering (Singer, Fulton, Davillier, Koshy et al., 2003) have also been related to lower levels of maternal distress.

#### **2.1.2.2.3. Extra-Familial Social Support**

Another major source of strength for both mothers and fathers of chronically ill children is the availability of support, primarily from extended family members. In the case of mothers, support from their own mothers and grandmothers has been found to be valuable (Oka and Ueda, 1998), and likewise for fathers (Pelchat et al., 2003). More than other family members and friends, grandparents have been found to be the most regular source of assistance for their children who have chronically ill children. It is suggested that through participation grandparents can better develop a relationship with the child with disabilities and get to know them on an intimate level. This helps parents feel proud of their child and can promote normalised attitudes within the family. As well as helping to alleviate physical exhaustion, assistance from grandparents also enables parents of chronically ill children to maintain a positive outlook (Green, 2001). The maintenance of a positive interpretation of one's child's situation has been related to a parent's success in receiving help from others (Ray,

2002). Indeed, parents who receive assistance from grandparents, also receive a greater number of other sources of support (Green, 2001). However, many parents do not like to ask for help from family members, particularly elderly parents who are perhaps unwell or affected by disability themselves, as they do not wish to saddle them with additional burdens (Green, 2001; Ray, 2002; Brewer et al., 2007). Equally, extended family members may not respond to one's child in the desired manner, may not be comfortable with disability, and may experience difficulty developing a relationship with the child. For some, extended family members 'just don't get it' and thus withdraw from visiting (Ray, 2002). Although this can be very hurtful for both parents, research has found fathers, in particular, to be disappointed by male family members' uneasiness with their child with disabilities (Pelchat et al., 2003). This supports other research that has found father's stress to be related to their child being accepted by others (Keller and Honig, 2004).

Social support is a topic that has been extensively studied over the years and one that has led to much debate about its relationship to stress and well-being (e.g. Hobfoll, 1988; Sarason, Sarason, Brock, and Pierce, 1996). The role that friends and family play in alleviating stress when a major life-changing event occurs remains to be unclear, although a wealth of evidence indicates that the support of others is beneficial to parents of children affected by chronic illness and disability (e.g. Warfield et al., 1999), and that perceived support has a positive impact on health and well-being (Schwarzer and Leppin, 1989). Equally however, the role that family and friends can play in exacerbating stress is little understood. In some cases parents can feel isolated and can experience the loss or breakdown of relationships when illness or disability becomes a part of their lives (Ray, 2002; Silver, Bauman and Weiss, 1999). Conversely, through the desire to protect their child (Speice, McDaniel, Rowley, and Loader, 2002) or because they feel that others don't understand their situation or feelings (Todd, 2002) they can isolate themselves and cut off access to family support. In other cases, family relationships can become more strongly cemented and new bonds can develop, as friends or family members commit whole-heartedly to the support of the parents and/or child (Ray, 2002; Green, 2001). However, receiving support is not necessarily synonymous with positive affect, as it has been related to feelings of guilt and dependency, and with lowered conscientiousness (Argyle, 1992). Nevertheless, *effective* support, whomever that may be from, and whatever form it

may take, is important to families of children affected by chronic illness or disability, as it empowers individuals and helps them to gain mastery over their lives (Rappaport, 1984). However, when considering what might constitute effective support, it is important to regard parents as individuals with different needs, as well as part of a family unit, both immediate and extended.

#### **2.1.2.2.4. Wider Social Support Systems**

Although having a child with chronic illness or disability does not inevitably lead to difficulties, many families commonly experience emotional, psychological, social, and political consequences, such as isolation and social marginalisation, stigmatisation, and disempowerment (Byrne, Cunningham, and Sloper, 1988; Pahl and Quine, 1987). One important protective factor for families affected by disability is support from larger social support networks (Wallander and Varni, 1998), which includes practical help and information as well as emotional support. Although there are agencies that can provide families with support such as medical advice and educational provision, no statutory provision exists which can encompass a family's complexity of needs (Hollins, 1985). Therefore, given that the nature of support from professional agencies is limited and that parents commonly experience social isolation, support from other parents in similar situations has significant potential to be helpful (Ray, 2002; Kerr and McIntosh, 2000; Oka and Ueda, 1998; Brown and Hepple, 1989; Holaday, 1994; McCubbin, 1984). Mutual support groups have many benefits, which include 'promoting a psychological sense of community, providing emotional support, providing role models, conveying a powerful ideology, providing information, offering ideas about ways of coping, giving the opportunity to help others, providing social companionship, and promoting a sense of mastery and control' (Solomon, Pistrang, and Baker, 2001:114). Being able to talk with other parents in similar situations has been found to be beneficial for coping (McCubbin, 1984).

Support groups can help parents by providing them with the opportunity to gather and share information with others who have 'been there', which increases understanding of the illness (Holaday, 1994). The experiential knowledge shared by people going through similar experiences can also enable parents to become active participants in



the management of their child's condition, providing them with a sense of confidence and control when faced with situations where they may previously have felt powerless to challenge unfavourable decisions (e.g. educational reviews, home adaptations, benefits, or health assessments) (Soloman et al., 2001). Support groups can offer parents a sense of belonging to a community, which can help them to feel valued and their feelings recognised. This can buffer feelings of isolation, stigmatisation, and social marginalisation (Soloman et al., 2001). Consequently, membership of such groups can lead to intrapersonal changes, including increased self-esteem and confidence, reduced guilt and self-blame, and greater acceptance of the child's disability. Such groups can also enable parents to view themselves in a more positive light and to ascribe more positive meaning to having a child with disabilities (Soloman et al., 2001). This can have a positive effect on parenting and in turn on children's behavioural problems, which consequently can increase parental sense of competence and self-efficacy (Solomon et al., 2001). Mutual support groups are not felt to be beneficial by all parents however, and some do not see association with other parents in similar situations as a source of support (Allan, Townley, and Phelan, 1974; Rawlins and Horner, 1988). Association with other families can painfully reflect one's own problems, and if a condition is progressive and incurable, this knowledge can be reinforced by seeing others in later stages of the disease (Coyne, 1997).

Social support can cover a range of phenomena (Sarason et al., 1996), which can include formal support from professionals, such as counsellors, health visitors, and doctors. Appropriate professional support can be helpful to parents of chronically ill children, making them feel listened to and taken seriously (Brewer et al., 2007). It can also help to reduce stress and increase coping by providing information and financial and emotional support (Sloper and Turner, 1992; Beresford, 1994). Aspects of professional support that are valued by parents include the willingness to try new things to improve families' situations, dedication and staying power, and flexibility and open-mindedness (Brewer et al., 2007). Other such support, which is intended to help, can be unhelpful however, and can cause significant distress to parents. Gaining the correct diagnosis for many chronic conditions can be a lengthy and painful process, which can lead to increased stress (e.g. Venters, 1981; Brewer et al., 2007). As a result parents can feel hostility and mistrust towards the medical profession (e.g.

Halliday, 1990). It is reported that medical staff can underestimate the devastation experienced by parents, providing them with little information and opportunity to discuss problems (Coyne, 1997). The way diagnosis is conveyed to parents can also be a moot point. If diagnostic information is given to parents in an unsatisfactory way, it can have implications for parents' later acceptance and ability to cope with the child's illness (Stein and Wooley, 1990). As well as with the medical profession parents can experience negative social interactions with other professional bodies and the public at large. This can include the criticism of parenting skills, failure to listen when parents are knowledgeable about a condition, failure to listen to explanations of the condition and having unrealistic expectations of the child, inflexibility in adapting to the condition, and instability and inconsistency of support provided by agencies due to staff turnover (Brewer et al., 2007).

While much social support is well-intended it is not always helpful (Suls, 1982), and it can end up being more of a stress than a support. It can be seen as a 'double edged sword', which can have a negative effect on well-being (Revenson, Schiaffino, Majerovitz, and Gibosky, 1991). Moreover, whether or not social support is deemed as helpful is a subjective judgement and subject to individual interpretations (Sarason et al., 1996). It is therefore important to understand parents' perceptions of support and establish which aspects of support are important for them. Parents' perceptions of support then require integration into work with families and their chronically ill children (Eiser, 1990).

### **2.1.2.3. Stress-Processing Resistance Factors**

No global interpretation of the care-giving experience can exist (Linge, 1976). While having a chronically ill child has been related to parent adjustment problems (e.g. Breslau et al., 1982; Nagy and Ungerer, 1990), many parents of chronically ill children have been found to function equally as well as parents of healthy children (e.g. Cadman, Rosenbaum, Boyle, and Offord, 1991; Spalding and Morgan, 1986). Each childhood chronic condition will pose its own set of unique challenges for the family, and parents are often put under considerable physical, emotional, and financial stress (Baldwin and Carlisle, 1994). However, this fact alone does not help in predicting how an individual parent will be affected (Olsson and Hwang, 2002). As

discussed, a number of intrapersonal and social-ecological factors play major roles in managing and buffering the stress presented by condition parameters (Wallander and Varni, 1992; Wallander et al., 1989). Further to this however, it is important to understand how the effects of stress are mediated by cognitive appraisal and coping processes (Lazarus and Folkman, 1984). Therefore, as well as the moderating effects of personal and social resources, individual appraisals of the situation are important to our understanding of the stress and coping process as they also serve as resistance factors (Wallander et al., 1989). The importance of stress processing factors like appraisal to parents' adjustment to having a chronically ill child has been highlighted by research. For example, the degree to which mothers' perceive the child's illness to impact on the family has been highlighted as a mediating process through which illness or functional severity may lead to parental psychological problems (e.g. Wallander et al., 1989; Ireys and Silver, 1996; Lustig et al., 1996). Mothers' appraisal of disability-related stress has also been strongly related to her maladjustment (Noojin and Wallander, 1997).

#### **2.1.2.3.1. Cognitive Appraisal and Coping Strategies**

The extent to which an event or situation is deemed stressful depends upon both the stressful stimulus and characteristics of the person experiencing it (Lazarus, 1999). Thus, it is the meaning that a stressful event holds for an individual that is vital in determining its psychological impact (Lazarus and Folkman, 1984). The appraisal of a stressful situation involves a transaction between external sources of stress and internal resources to cope with them. The stress reaction is elicited if the individual appraises the situation as insurmountable. The differential ways in which people appraise stress is related to their subsequent coping efforts. The way in which individuals respond to stressful situations then depends upon a number of internal and external resources and moderating factors as discussed (Lazarus, 1966; Lazarus and Folkman, 1984). It is therefore not necessarily the stressor alone that has a consequence for an individual or family, but the combination of the stressor and the person's view of him or herself and the world around them. These views contribute to make the stressors comprehensible to the individual who experiences them (Olsson and Hwang, 2002:549), and has been conceptualised as a Sense of Coherence (SOC) (Antonovsky, 1987). Sense of Coherence is defined as a:

‘global orientation that expresses the extent to which one has a pervasive, enduring though dynamic feeling of confidence that: (i) the stimuli deriving from one’s internal and external environments in the course of living are structured, predictable, and explicable; (2) the resources are available to meet the demands posed by these stimuli; and (3) these demands are challenges, worthy of investment and engagement and that life makes sense emotionally’ (Antonovsky, 1987:19).

These three components are referred to as *comprehensibility*, *manageability*, and *meaningfulness*. It is proposed that a strong SOC enables a person to define a stressful situation as non-stressful (Antonovsky, 1987). A person with a strong SOC will perceive a situation as a challenge rather than a threat, have confidence that things will work out, and believe they have the resources to cope and that all will become clear and manageable. Aspects of SOC such as *meaningfulness* can be enhanced by having a chronically ill child if the experience is perceived as satisfying and a positive challenge. *Comprehensibility* can be strengthened by increased knowledge about a condition, and *manageability* can be improved by programmes that assist parents in the management of child behavioural problems, for example (Beresford, 1996; Bristol, Gallagher and Holt, 1993). However, while some consider SOC to be a stable trait (e.g. Antonovsky, 1993; Schnyder, Buchi, Sensky and Klaghofer, 2000), others purport its potential to be moderated by chronic stressors (Antonovsky, 1993). Thus, a chronic stressor like having a chronically ill child can have a potentially negative impact on an individual’s SOC, making them vulnerable to further stress and distress (Olsson and Hwang, 2002). Low SOC has been strongly linked to depression (Flannery, Perry, Penk and Flannery, 1994).

Coping processes are purported to have two parallel functions. These relate to the regulation of emotional control, and problem-solving or functional coping (Lazarus and Folkman, 1984; Moos and Schaefer, 1986; Rutter, 1981), and describe parents’ emotional states and their ability to function in their parenting and family roles (Melnik et al., 2001). The appraisal of one’s cognitive and behavioural efforts to meet the demands that childhood chronic illness present, both internally and externally, determines how one feels about the situation and one’s ability to physically manage.

Indeed, the emotional well-being of mothers of chronically ill children has been linked to their perceptions of the situation in which they find themselves (Tunali and Power, 1993). While appraisals precede coping efforts (Lazarus and Folkman, 1984), coping can also be seen as a mediator between illness representations and outcomes (Leventhal et al., 1997). Positive appraisal of a coping strategy can lead to representations of an illness being modified and seen in a more positive light, and in turn to improved feelings about the situation and one's ability to handle it. Healthy cognitive appraisals are therefore vital resistance factors in parents' adaptation (Wallander et al., 1989), and the way in which families make sense of the illness the child is affected by is fundamental to adaptation.

The ways in which individuals cope however can either be active or avoidant in nature (Billings and Moos, 1981), which has different implications for adjustment as it ostensibly moderates the effects of illness-related stressors on adjustment (Wallander et al., 1989). Avoidant coping strategies are described as behaviours and cognitions that are intended to distract attention away from the stressful situation. Cognitive avoidance or denial is one form of avoidant coping, which can be adaptive in the short term by allowing an individual time to organise and gather resources. However, if continued to be relied upon this form of coping can result in psychological dysfunction (Moos and Schaefer, 1984) and it is seen as a significant risk factor to adaptation (Vaillant, 1977) and has been associated with poor adjustment in both children and adults even in healthy families (e.g. Holahan and Moos, 1987; Moos, Brennan, Fondacaro, and Moos, 1990). Active coping on the other hand describes strategies that directly affect the stressor by either minimising or eliminating it. This can be done behaviourally or cognitively by interpreting the stressor in a more positive light (Billings and Moos, 1981). Active coping strategies such as the establishment of routines has been associated with better adjustment amongst mothers of chronically ill children as discussed (e.g. Deatrick et al., 1988; Gibson, 1988; Knafl et al., 1996), as has the positive interpretation of the situation (McCubbin and McCubbin, 1993), and the use of cognitive restructuring techniques (Hill, 1994; Ray, 2002). As previously stated, the meaning of a stressful event plays a vital role in determining its psychological impact (Lazarus and Folkman, 1984).

#### **2.1.2.3.2. Assigning Meaning to the Illness**

Although adjustment to chronic illness is often associated with negative affect and emotional distress, many people have positive experiences. Adjustment to chronic illness can provide individuals and families with a ‘meaningful event’, which increases their value in life and relationships (Folkman, 1997; Wright and Kirby, 1999). Many parents of chronically ill or disabled children have recognised the development of special qualities and strengths in themselves, their children, and other family members (Hill, 1994; Eiser, 1994), and reported that the experience has provided the family with the opportunity for growth (Summers, Behr and Turnbull, 1989). The ability to define illness as a meaningful experience has been related to better family adjustment (McCubbin and McCubbin, 1993). Conversely, the negative definition of the situation has been found to be ‘the single most important predictor of parental stress’ when caring for a child with disability (Saloviita et al., 2003:300). People who are able to put things in a religious context have also been found to cope better than those who cannot (Venters, 1981). In the situation where a child has survived against all adversity, families can give special meaning to the child’s place in the world and believe their survival to be fated (Mayer, 1982). Religion is a key resource for some parents as prayers help them to cope with manifestations of the illness and enable them to view the experience of having a chronically ill child as enhancing their own spiritual growth (Hill, 1994; Williams, 1993; Kelleher and Islam, 1996).

Families of chronically ill children interpret their situations in a number of ways as they adjust to new and changing experiences and learn to cope. In some instances parents use positive reframing and the optimistic definition of events as coping strategies (Hill, 1994), which have been found to contribute to the maintenance of family unity (Venters, 1981; McCubbin, 1984). For example, downward comparisons can be made between the chronically ill child and children whom parents consider to be worse off, thus considering the ill child or their family to be ‘lucky’ (Affleck and Tennen, 1993; Ray, 2002). Downward comparison (Festinger, 1957) is a form of emotion-focused coping, which it is suggested may be used in order to improve self-esteem (Taylor, Lictman, and Wood, 1984). It has been observed as a coping strategy

in those with other chronic conditions, such as childhood multiple sclerosis (Boyd and MacMillan, 2005). Other examples of positive reframing in this context include the emphasis being given to small achievements or developments the child has made, thus perceiving the child as strong, in control, and a fighter (Ray, 2002), and parents considering their child to have abilities that compensate for limitations (Hill, 1994). Normalisation is another coping strategy that parents of chronically ill children often employ (Eiser, 1990; Monsen, 1999). It is aimed at minimising stigma and creating a sense of control (Eiser, 1990). This can include attributing symptoms to non-disease related causes, making favourable comparisons between their child and other children who are not chronically ill, and separating the 'normal' aspects of a child's life from the illness-related aspects (Eiser, 1990; Hill, 1994; Monsen, 1999).

Drawing on inner strength, making the best of a situation, and being positive has been found to contribute to maternal confidence and feelings of empowerment (Gibson, 1995; Sullivan-Boyai et al., 2003). More negative appraisal of the situation on the other hand can have a significant impact on parents' ability to effectively cope. A number of emotional distress factors have also been highlighted as being particularly salient in both acute and chronic childhood illness situations, and are suggested to interfere with healthy cognitive appraisal, invading illness representations and perceived self-efficacy (Bonner, Hardy, Guill, McLaughlin et al., 2006). A number of emotional distress constructs have been examined in relation to parent adjustment and their psychological management of a childhood chronic condition.

#### **2.1.2.3.3.           Uncertainty and Perceptions of Child Vulnerability**

One factor that can hinder the coping process is uncertainty. Uncertainty is conceptualised as a cognitive state which affects parent adjustment as a result of the pervasive fear of possible illness consequences (Stewart and Michel, 2000). Anxious cognitions about child susceptibility to future illness are also referred to as perceptions of child vulnerability (Forsyth, Horwitz, Leventhal, Burger et al., 1996), and a number of studies have highlighted the relationship between uncertainty and psychological distress (Stewart and Mishel, 2000). It is considered to be a multidimensional construct (Cohen, 1995; Mishel, 1991, 1999). Among parents of chronically ill children a number of dimensions of uncertainty have been identified,

including uncertainty about the illness event, the time frame or course of the illness, the aetiology of the illness, the treatment, and the prognosis (Cohen, 1993b). Uncertainty regarding the child's life expectancy and the predictability of daily symptoms are two other dimensions that have been explored in the childhood chronic illness literature (Dodgson et al., 2000). Uncertainty can pervade throughout the child's illness having a significant impact on parent's psychological management of the condition (Mishel, 1983). It is a major factor influencing expectations about treatment and prognosis (Hilton, 1992) and invades all stages of the condition (Swallow and Jacoby, 2001). Parents can experience uncertainty in both acute and long-term contexts. Acute uncertainty pertains to episodes such as decision-making regarding treatment, and long-term uncertainty presents parents with concerns about the child's future well-being and dilemmas about the implications of past decisions or events on the child's long term health and achievement (Mishel, 1981; Bonner et al., 2006).

Uncertainty can also lead to further emotional responses such as grief (Gibson, 1995), guilt and worry, and unresolved sorrow and anger (Bonner et al., 2006), which not only have implications for parents' adjustment but also for the psychosocial development of the child (Thompson and Gustafson, 1996). Guilt and worry are seen to be related to parents' fearfulness about parenting, their ability to manage acute episodes of a condition, and making decisions for a very ill child. It has been positively related to perceived impact on the family and parenting worries, and it is purported that parents who score highly on this factor perceive their child as fragile (Bonner et al., 2006). This has implications for parents' cognitive appraisal of internal resources and self-efficacy, which are important variables to include when evaluating the factors that contribute to family adjustment (e.g. Kazak et al., 2004). Another element of parent distress that has been identified as important to the appraisal process is unresolved sorrow and anger, which pertains to ongoing feelings of loss associated with the child's illness, such as the loss of a healthy child and of a perceptually easy and normal life course (Bonner et al., 2006). Unresolved sorrow and anger has been positively related to perceived impact on the family, depression, and increased levels of emotional strain caused by, and intrusive thoughts about, the child's illness (Bonner et al., 1996). This element of parent distress may therefore



indicate less adaptive functioning, which it is suggested may impact upon child adjustment (Thompson and Gustafson, 1996).

The impact that certain parent cognitions can have on child adjustment has not been widely studied. One type of cognition that may relate to child adjustment however, is parental perceptions of child vulnerability. The term 'perceptions of child vulnerability' was first used by Green and Solnit (1964) to describe parental anxiety about their child's health. They proposed that such anxiety brought about maladaptive patterns of parent-child interaction and child behaviour problems called 'a vulnerable child syndrome' (Estroff, Yando, Burke, and Snyder, 1994; Leslie and Boyce, 1996). Increased perceptions of child vulnerability and parental anxiety about the child's susceptibility to illness have been linked to a number of outcomes for children that could interfere with their adjustment, including increased absence from school (Spurrier, Sawyer, Staugas, Martin et al., 2000), increased health care use (Maiman, Becker and Katlic, 1986; Fiegelman, Duggan, Bazell, Baumgardner et al., 1990), increased social anxiety (Anthony et al., 2003) and an increase in child dependence, demandingness, and behavioural difficulties (Bendell, Field, Yando, Lang et al., 1994). Higher perceived child vulnerability has also been related to more negative outcomes for the child in terms of adaptive functioning (Allen, Manuel, Legault, Naughton et al., 2004). Such parental cognitions may impact on child adjustment by interfering with the critical development of behavioural autonomy through over-involvement with the child and the adoption of an over-protective parenting style (Anderson and Coyne, 1991; Holmbeck, Johnson, Willls, McKernon et al., 2002).

When a child is perceived as vulnerable, it is a parent's natural instinct to want to protect them (Lollar, 1994). However, mothers are more likely than fathers to be over-protective, and are at more risk of becoming engulfed by the role they play in the care of their chronically ill children (Pelchat et al., 2003). In such a situation the caring role becomes the defining feature of self-identity, and it is difficult for mothers to separate themselves from the caring role. Mothers can also identify so strongly with their children on an emotional level that they personally experience their child's pain and suffering (Twigg and Atkin, 1994; James, 1998). Their role as parent in the child's adaptation is thus seen as the dominant issue in mothers' lives (Pelchat et al., 1999a, 1999b), and having one's child diagnosed with a chronic illness can throw

meaning and order into disarray (Williams, 1984). Childhood chronic illness can thus create a 'biographical disruption' for mothers as it transforms the mother's role from that of mother of a healthy child to that of mother of a 'child in crisis' (Bury, 1982). Such biographical disruption can also have implications for paternal adjustment and for family functioning.

#### **2.1.2.3.4. Childhood Chronic Illness as Biographical Disruption**

Childhood illness is conceptualised as representing a 'condensed symbol of childhood itself through intensification of concepts of dependency and vulnerability' (James, 1998:97). This can impact on the family long-term by altering parents' biographies and changing the way in which a family functions. As previously discussed, the different ways in which mothers and fathers appraise stressful life events is related to the gender roles assigned to men and women in our culture (Gray, 2003). From this perspective one can consider gender roles as having their own set of biographies, which determine what happens when throughout the life cycle according to societal norms. The arrival of a chronically ill or disabled child therefore disrupts parents' biographies and causes psychological distress. However the focus of the distress is determined by the gender role assumed by the parent. For example, it is purported that mothers are more likely than fathers to blame themselves when their child has difficulties and to have their identities threatened when their children are affected by illness or disability (Anderson and Elfert, 1989).

It is suggested that mothers' identities are threatened by illness and disability in their children because it is their role to nurture and protect them, and to care for their emotional and physical well-being (Bury, 1982; Young, Dixon-Woods, Findlay, Heney et al., 2002). This in turn takes a physical and emotional toll on the mother. When a child is diagnosed with a chronic condition mothers have reported feeling a part of their child, having to endure the sight of the child in pain and distress, feeling the child's feelings, and wishing they could take the child's place. It is also purported that part of the mother's role is to keep the child occupied and the situation light hearted and fun as a means of staving off distress and shielding the child from the reality of the situation (Bury, 1982). Ostensibly, mothers also have to develop the new role of 'information broker', where information is filtered and censored accordingly

(James, 1989). Further in her role as protector, the mother also helps her child to 'pass as normal' (Goffman, 1968), by way of protecting the child's self-identity and wanting them to be recognised for both their individuality and their sameness (Landsman, 1988). However, whilst encouraging others to treat her child no differently from others, new forms of relationship have to be negotiated, since normal parenting and discipline strategies are disrupted and deemed no longer appropriate by the diagnosis of life threatening conditions (Young et al., 2002).

While mothers' concerns are suggested to be ensconced in their nurturing and protective parenting role, fathers' expectations regarding their chronically ill or disabled child are professed to be related to the world outside the family (Pelchat et al., 2003). Since it is the father's role to prepare his children for their future in a more practical and industrial sense, their anxiety tends to centre around their child's future occupational achievement and long-term social status. Fathers' concern for their child's future social and economic potential is suggested to be exacerbated when the child's disabilities are marginal, thus making expectations unclear (Trute, 1995), and men have been found to be more prone to depression if their ill child is male (Trute, 1995; Frey, Greenberg, and Fewel, 1989; Tallman, 1965). They have also been found to be disturbed by their child's disability (Pelchat et al., 2001b), and have been reported by mothers as being uncomfortable with their child's condition and reluctant to ask others for help or advice (Pelchat et al., 2003). However, this is purported as being a reaction to possible stigmatisation as generated by the disability rather than a reaction to the disability itself (Pelchat et al., 2003), and is perhaps related to their concern that their child is accepted in the wider world (Keller and Honig, 2004). It is suggested that fathers want their child to be thought of as being 'normal', and prefer others to notice the similarities rather than the differences between their child and other children. This concern, it is suggested, surrounds the possibility that their fatherhood or social status could be called into question (Parent, 1992).

Despite the different ways in which mothers and fathers respond to childhood chronic illness, adaptation for both is an ongoing process whereby their lives can be taken over by the experiences and needs of their child. Despite the difficulties experienced however, both mothers and fathers have described the enriching nature of the experience of having a child affected by a chronic condition and how they do their

utmost to adapt to their child's needs: "after all that has taken place we've realised that if we had not had our child, we would have missed something" (Pelchat et al., 2003:242). This illustrates how parents try to restore a sense of meaning and order into their lives by reconstructing their biographical narratives (Williams, 1984). As discussed, although there are many experiences which parents of chronically ill and disabled children share, they may do so differentially, and childhood chronic illness may hold different meaning for mothers and fathers. Their perception of the situation and appraisal of their ability to source and use resources to cope may also differ. Mothers' and fathers' expectations of one another in their parenting roles and their ability to give appropriate support is therefore important to parent coping, and in turn to family functioning and child adaptation (Pelchat et al., 2003).

As discussed, families can draw on a plethora of internal and external resources, the suitability and availability of which are subject to change over time as the functioning of the family and the needs of the individual shift. Stress and coping processes are therefore dynamic and ongoing and subject to a series of ups and downs, and perceptions of the illness and one's situation is subject to repeated modification (Wortman and Silver, 1987; McCubbin and McCubbin, 1991; Leventhal et al., 1997). Since family functioning has been consistently identified as a predictor of psychological functioning of children with chronic illness (e.g. Finney and Bonner, 1992; Quittner and Digirolamo, 1998; Wallander et al., 1989) an understanding of the variables that are associated with it is essential. Furthermore, as previously mentioned, recent research has demonstrated the importance of the father- as well as the mother-child relationship in child psychosocial outcomes (Rohner and Veneziano, 2001). The ways in which these relationships can be disrupted by illness or disability could have a profound effect on how children adjust psychosocially.

## **2.2. Living with Chronic Illness and Disability: The Impact on the Child**

Children with disabilities or chronic illnesses have been shown to be at increased risk for psychological morbidity and adjustment difficulties (e.g. Lavigne and Faier-Routman, 1992; Bennett, 1994; Noll, Gartstein, Vannatta, Correll et al., 1999; Koopmans and Lamers, 2000). Indeed, a wide range of psychosocial problems has been associated with chronic illness including anxiety, depression and adjustment disorders (Pitts, 1991; Lavigne and Faier-Routman, 1992; Koopmans and Lamers, 2000), and problems in peer relationships (e.g. Noll, LeRoy, Bukowski, Rogosch et al., 1991). Studies exploring different childhood disabilities have shown similarities in the psychosocial problems associated with them (Stein and Jessop, 1989; Perrin and MacLean, 1988), although few have been able to directly relate condition type (Thompson and Gustafson, 1996) or disease severity (Wallander and Varni, 1998) with children's psychosocial adjustment. The literature on psychosocial adjustment and childhood chronic illness and disability is by no means consistent however, as other studies have found children with chronic illness to be well adjusted psychosocially and to not differ greatly from their healthy peers in terms of mental health problems or achievements in developmental milestones (e.g. Ievers and Drotar, 1996).

Depending on the condition and its severity, children and young people affected by chronic illness are faced with a multitude of potential difficulties including a variety of possible medical symptoms and disease characteristics and unpredictability, which is characteristic of many chronic conditions. Children with chronic illness are also challenged to cope with painful physical episodes, hospitalisation, and time away from their families, and to often endure complex treatment regimens that require them to negotiate with their parents to manage the condition. This can cause conflict and disrupt family routines (e.g. Delambo et al., 2004; Ievers and Drotar, 1996). Chronic illness can also force children to have increased school absences and to be limited in their social activities both in and outside of school (e.g. Wallander and Varni, 1998). However, while studies show compelling commonalities in the psychosocial problems associated with a range of different childhood disabilities (e.g. Perrin and MacLean, 1988), most studies show no evidence that disease severity affects adjustment

(Wallander and Varni, 1998), and as with family adaptation the adjustment of children and young people with chronic conditions has been found to be more related to parent, child, and family factors than to disease or disability parameters (Lavigne and Faier-Routman, 1992).

The following section will explore illness-related and psychosocial factors that put chronically ill children and adolescents at risk for adjustment difficulties in line with Wallander and colleagues disability stress-coping model (Wallander et al., 1989; Wallander and Varni, 1992). Also in line with this model, the role that protective intrapersonal, social-ecological, and stress processing factors play in buffering the impact stressors have on adaptation will also be explored. Since there is no literature that describes the psychosocial adjustment of children and young people living with MPS IH post-BMT, it will be necessary to draw on research that has explored child and adolescent adjustment to other chronic, life-limiting, and disabling conditions in order to examine the risk and resistance factors that may be relevant to this patient group. Chronic illnesses that will be discussed will include sickle cell disease, cystic fibrosis, juvenile rheumatoid arthritis, and diabetes. Since children affected by MPS IH post-BMT also have short stature and experience significant limitations to their physical, cognitive, and adaptive functioning, disabilities that have similar manifestations will also be discussed.

### **2.2.1. Risk Factors**

#### **2.2.1.1. Illness Related Risk Factors**

Many chronic illnesses present with symptoms that can result in discernable distress for children and adolescents, as can the associated treatment regimens. Young people can have fears about the course of a condition and the implications it has for their future (e.g. Barlow, Harrison, and Shaw, 1998). This can be particularly distressing when a condition is life-limiting (e.g. Ievers and Drotar, 1996; Odegard, 2005). Many conditions cause the child to experience pain and physical limitations, life-threatening complications, hospitalisations, and surgery (e.g. Cassidy and Petty, 1995; Burlew et al., 2000). Physical and drug therapies, and diet management (e.g. Cassidy and Petty,

1995; Stark, Jelalian, and Miller, 1995; Boland and Grey, 2000) are also often required, which can be frustrating, laborious and time consuming. These can also disrupt family routines and can put and parent-child relationships under strain (DeLambo et al., 2004). Young people can also find balancing the management of normative and illness-specific caretaking responsibilities difficult (e.g. Ievers and Drotar, 1996) and can find adherence to treatment regimens problematic, particularly in adolescence (e.g. Gudas, Koocher, and Wypij, 1991; Hauser Jacobson, Lavori, Wolfdorf et al., 1990). This can have serious implications for health and well-being (Patterson, Budd, Goetz, and Warwick, 1993). Children and young people with chronic illnesses can also often miss school due to illness-related complications, exacerbations, and hospitalisations (Gil, Porter, Ready, Workman et al., 2000), and may miss out on the opportunity to participate in social activities because of illness-related restrictions (e.g. Wallander and Varni, 1998). They may also experience altered body image, anxiety around teasing and social acceptance (Brown et al., 1993; Barlow et al., 1998; LaGreca, Prinstein, and Fetter, 2001), and limitations in perceived academic, social, and athletic competence (e.g. Telfair, 1994; Casey, Brown, and Bakeman, 2000).

In recognition of the stressors and burdens associated with many chronic conditions, a considerable number of studies have been conducted to assess the impact the illness has on children's psychological adjustment. However, a consensus about the psychological impact of many conditions has not been reached. For example, in the case of chronic arthritis, some authors report higher rates of adjustment problems (e.g. behavioural and emotional problems, internalising problems, low self-esteem and poor self-concept) in relation to norms (e.g. Timko et al., 1992; LeBovidge, Lavigne, Donenberg, and Miller, 2003), while others report no significant differences (e.g. Noll et al., 2000). Sickle cell disease is also posited to be a risk factor for adaptation (e.g. Bennet, 1994; Thompson et al., 1999). However, adaptation has been found to vary due to fluctuating symptoms (Thompson et al., 1993; Thompson., Gil, Keith, Gustafson et al., 1994). Further findings are inconsistent regarding whether or not adolescents affected by sickle cell disease differ from peers on depression and other internalising disorders (e.g. Brown, Kaslow, Doepke, Buchanan, et al., 1993), anxiety (e.g. Schoenherr, Brown, Baldwin, and Kaslow, 1992), externalising disorders (Brown, Doepke et al., 1993) and social relations (Noll, Bukowski, Davies, Koontz et

al., 1992). Thus, while chronic illness is proposed as a risk factor for adjustment, there is little consensus that biomedical and illness-related factors alone contribute to adjustment problems (Lavigne and Faier-Routman, 1992).

Again with regard to functional status, the literature is inconsistent regarding the relationship between the ability to perform everyday tasks appropriate for a particular age and children's psychosocial adjustment (Dadds, Stein and Silver, 1995; Lavigne and Faier-Routmen, 1993). Some studies have found functional status to affect adjustment (Stein and Jessop, 1984; Heller, Rafman, Zvagulis and Pless, 1985; Witt et al., 2003) while others have not found such a relationship (Padur, Rapoff, Houston, Barnard et al., 1995). Of those studies that have found a relationship between adjustment and functional status, children with learning and communication impairments have been found to experience poor psychosocial adjustment, however this was not related to self-care and mobility impairments (Witt et al., 2003), lower adaptive and developmental competencies have been related to maladjustment amongst children affected by sickle cell disease (Hurtig and Park, 1989; Casey et al., 2000), and adolescents with progressive physical disability have been reported as becoming increasingly socially withdrawn as physical status declines (Harper, 1983). Furthermore, in conditions that present with delays in growth and limitations in physical activities, boys have been found to experience greater adjustment difficulties than girls (e.g. Hurtig and Park, 1989; Casey et al., 2000), though this is not a consistent finding. As with family adaptation however, a wealth of evidence suggests that biomedical factors alone account for little of the variance in adaptation and that psychosocial factors are more influential (Bennet, 1994; Lavigne and Faier-Routman, 1993; Thompson et al., 1999). While chronic illness is recognised as a risk factor for adjustment, individual and family factors have been shown to have a moderating effect on these stressors (Wallander et al., 1989).

#### **2.2.1.2. Psychosocial Risk Factors**

Having a chronic illness during childhood can substantially interfere with day-to-day functioning, and the social experiences of chronically ill children and adolescents often differ from those of their healthy peers. Absences from school due to hospitalisation or doctors' visits may interfere with an individual's inclusion in school



activities and opportunities for socialisation with peers. Aspects of the condition itself may also limit an individual in their ability to physically 'keep up' with peers or participate in age-appropriate social activities, and the way an individual feels about themselves and their self-image may hinder their willingness to socialise (Charache, Lubin and Reid, 1989; Wasserman et al., 1991; Telfair, 1994; Kapp-Simon, 1997). Issues of independence may further limit opportunities to participate in activities with peers outside of school (McAnarney, 1985). Chronically ill children can therefore experience isolation, emotional distress, and lack the opportunity to develop social skills (e.g. Telfair, 1994). While the presence of physical illness does not necessarily impact upon peer acceptance (Guite, Walker, Smith, and Garber, 2000), conditions that present with visible features and affect physical appearance or limit physical activities have been associated with peer relationship difficulties (Schuman and LaGreca, 1999; Wray, Long, Radley-Smith, and Yacoub, 2001). Children affected by such conditions have been found to experience lower social acceptance than their peers (Schuman and LaGreca, 1999) and to be more reluctant to initiate and engage in peer relations (Kapp-Simon and McGuire, 1997).

As children grow older and become aware of physical limitations and differences between themselves and other children, issues may arise for some. This usually happens during the cognitive stage of concrete operations (age 7-11 years) as children begin to gain an understanding of how the body functions and about their illness and its causality, and the implications of treatment regimens (Lewis and Vitulano, 2003). Children with short stature have reported feelings of vulnerability and humiliation in relation to their physical status, which have in turn been associated with social and behavioural problems, and cognitive and social development (Frankel, 1996). Older children and adolescents with chronic conditions have also reported lower perceived athletic and social competence than their healthy peers (King et al., 1993), lower perceived peer acceptance and competence (Scott and Scott, 1999), and they have reported others to see them as different and to not have good peer relationships (Skar, 2003). While young children with chronic illness are generally found to adjust well emotionally and socially (e.g. Ievers and Drotar, 1996) and to feel they have social competencies similar to their peers (e.g. Lemanek, Horwitz, and Ohene-Frempong, 1994; Brown et al., 1993), adolescents can experience difficulties associated with body image, social activities, and adjustment (e.g. Charache et al., 1989; Brown et al.,

1993). Adolescence can therefore be difficult for chronically ill or disabled individuals, as it is a time of significant physical and psychological change which can be disrupted by chronic illness.

Early adolescence (10-14 years of age) is a time of pubertal growth, identity formation, independence, and peer group development. There is much same-sex behaviour where individuals strive to fit in and adopt an identity that is in keeping with peer groups. This is a time of asking '*who am I in this changing body?*' Adolescence is also a time when an element of independence from the family is achieved, which should include taking responsibility for self-care tasks and taking part in family responsibilities, such as household chores. The transition from childhood to adolescence is a crucial time in terms of behavioural autonomy, and by this time decision-making responsibilities should have shifted from parent to child (Anderson and Coyne, 1991). If this transition is disrupted by disability, ill health, or parenting, familial, or social factors, behavioural problems may ensue, taking the form of both externalising and internalising behaviour, such as depression and withdrawal (Anderson and Coyne, 1991). Furthermore, if delays in psychosocial development occur in early adolescence, it is likely that further delays will be experienced throughout the remainder of adolescence and chronically ill adolescents can fail to reach adult social maturity (McAnarney, 1985). Forced dependence on the family as a result of the illness can therefore challenge adolescents' strivings for independence from the family (McAnarney, 1985). Adolescents and young adults chronically ill since birth have been found to exhibit more psychosocial problems associated with future plans than young people not affected by chronic illness (Orr et al., 1984). The implication here is that chronically ill young adults can fail to make the transition from middle to late adolescence and a more future orientated way of thinking, and can therefore remain attached to the family. This has been evidenced by research, as young adults affected by childhood chronic illnesses have more often been found to be unmarried and living at home with their parents than controls. They have also been found to experience difficulty in separating from their parents, to have poor social maturation, delayed sexual development, and to live a more dependent lifestyle (Kokkonen, Lautala, and Salmela, 1993; Kokkonen, 1995).

#### **2.2.1.2.1.        Learned Helplessness and the Sick Role**

A process that is key to understanding chronically ill children's increased dependence on the family and psychosocial difficulties is learned helplessness. Learned helplessness is defined as 'an uninformed process of withdrawing from definable, manageable adversities by succumbing to everyday problems' (Braden, 1990:42). The development of such a response has been associated with adjustment difficulties (Braden, 1990) and is suggested as having implications for cognitive functioning (Thompson et al., 2002). Although some aspects of an individual's response to chronic illness are believed to be mediated by certain personality traits (Kobasa, 1979), a large part of the response is purported to be learned (Braden, 1990), and it is contended that an improved quality of life can be achieved if an individual acquires skills that will enable him or her to be more resourceful and to carry out self-help activities (Braden, 1990; Rosenbaum, 1983). Indeed, decision-making and household responsibilities have been related to functional status, self-management, and social competence in young people with Spina Bifida (Sawin, Buran, Brei and Fastenau, 2003), self-reliance and adaptive competence have been related to adjustment in children with sickle cell disease (Hurtig and White, 1986), and competitiveness in employment has been positively related to adaptive skills in young people with learning disability (Stephens, Collins and Dodder, 2005).

This supports the importance of the development of autonomous behaviour throughout childhood and adolescence (Anderson and Coyne, 1991), and suggests that employment itself may enhance the adaptive skills of people with developmental disability (Stephens et al., 2005). These issues are also particularly pertinent to individuals who have been affected by chronic illness since birth, where depending on the family environment and parental attitudes, the child would have grown up learning to live with illness and disability in a particular way. Moreover, it is possible that an element of an individual's identity that has been affected by a condition since birth may be ensconced in the actual medical condition the child is affected by, or in 'being ill' or 'being disabled' (Royer, 1998). There is argument that 'being sick' constitutes a social role, and in some instances the 'sick role' can bring with it privileges and exemptions, which may encourage an individual to perpetuate that role (Parsons, 1951).

Learned helplessness is also a factor that contributes to the psychosocial development of children with learning difficulties (e.g. Arnold, Goldston, Walsh, Reboussin et al., 2005; Sundheim and Voeller, 2004). Children that have early experiences of reading failure when faced with words they are unfamiliar with have been found to experience feelings of defeat and helplessness (Cirorowski, 1992). This is then purported to lead to the failure to develop adequate reading strategies, and consequently to a lack of interest in furthering the reading experience. Additionally, when children with learning difficulties experience failure in the classroom, it has implications for their self-esteem and for other areas of their life outside of the school setting (Hersh, Stone and Ford, 1996). Thus, when an individual experiences failure in an academic setting, social performance may be affected as well, rendering an individual inhibited, withdrawn, and less proactive. Research has found children with learning difficulties to be at increased risk for social (Pihl and McLarnon, 1984), behavioural and emotional (Dean, 1985), and academic difficulties (Cirorowski, 1992). In terms of academic skills, a number of emotional and cognitive-motivational factors are at play to influence the performance of a child. Academic failure, low self-concept, and low motivation, which is further exacerbated by anxiety about further failure all contribute to an individual's lower achievement in an academic setting (Hersh et al, 1996).

Moreover, the effect that academic failure has on motivation and self-concept has enormous implications for a child's psychosocial development, as the process involves the expectation of failure, lowered motivation, perceptions of personal inadequacy, and lack of persistence, which pervade into other areas of an individual's life (e.g. Dean, 1985; Friedman and Medway, 1987; Garber and Seligman, 1980; Licht and Kistner, 1986). This can then have an effect on an individual's opportunities to improve their academic potential, and can create a vicious circle, as social and behavioural problems have been found to mask an individual's learning ability. Some children and young people affected by chronic illness who have mild learning difficulties are said to go to special needs schools quite unnecessarily, as a result of social, behavioural, and emotional difficulties, which have masked their academic potential (Whittington, Holland, Webb, Butler et al., 2004). This is essentially an

illustration of learned helplessness (Seligman and Maier, 1967), which may be particularly pertinent to adolescents.

#### **2.2.1.2.2. Parenting and Family Risk Factors**

A number of parental and family related factors have been associated with the psychosocial development of children and young people affected by chronic illness or disability. This has implications not only for the development of an individual's internal coping mechanisms, but also for cognitive development and the development of adaptive, resourceful, and self-help skills. For example, low parental expectations of the child and the fostering of dependence have been found to have a detrimental effect on the psychosocial adjustment of young people with disabilities and chronic illness (McAnarney, 1985; Orr et al., 1984). Mothers' childrearing behaviours have also been linked to child cognitive function. These include nurturing behaviours, parent-child interaction (Kolobe, 2004) and stimulation (Armor, 2003), parent distress (Singer, Fulton, Davillier, Koshy et al., 2003), and parental risk in terms of a learned helplessness attributional style (Thompson et al., 2002). Poor family functioning (Drotar, 1997), low income (Gortmaker, Walker, Weitzman and Sobol, 1990; Brown et al., 1993), high family control (Mink, Meyers, and Nihira, 1984), maternal distress (Lavigne and Faier-Routman, 1993; Thompson et al., 2002), post-diagnosis maternal adjustment (Sawyer et al., 1998), and the burden that illness places on the family (Daniels et al., 1987) are further family-related factors that have been associated with chronically ill children's psychosocial maladjustment. Parenting and family factors have also been found to impact on the chronically ill child's health status (e.g. DeLambo, Ievers-Landis, Drotar, and Quittner, 2004), and create feelings of uncertainty about the illness (Mullins, Wolfe-Christensen, Hoff Pai, Carpentier et al., 2007), which can lead to the risk of depression and feeling out of control of one's life (Brown, Armstrong, and Eckman., 1993).

As previously discussed, mothers of chronically ill children can become ensconced in their parenting role, over-involved with their child, and prone to employing an over-protective parenting style (e.g. Bury, 1992; Pelchat et al., 2003; Twigg and Atkin, 1994). This has been found to have a detrimental effect on children's adjustment, as the early development of internal resources such as behavioural autonomy can be

disrupted (Holmbeck et al., 2002). Numerous studies have found associations between parental over-protectiveness and a range of emotional and behavioural difficulties, such as depression and externalising behaviour (e.g. Cappelli et al., 1989; Burbach, Kashani and Rosenberg, 1989), and behavioural autonomy is considered to be the mediating factor between parental over-protectiveness and child psychosocial adjustment (Holmbeck et al., 2002). Parental anxiety and perceptions of child vulnerability have also been related to child internalising problems (Estroff et al., 1994), increased social anxiety in children (Anthony et al., 2003), young people's perceptions of illness uncertainty (Mullins et al., 2007), and illness-related anxiety and the development of anxious attachments to parents (Odegard, 2005). The way that families function around and react to childhood chronic illness has significant implications for children's coping. It influences the way in which young people appraise their illness and the situation they are in and their ability to cope, and impacts upon coping resources such as health control beliefs, self-reliance, and self-efficacy, which are important to young people's adjustment (e.g. Lau, 1982; Hurtig and White, 1986; Helgeson, 1992; Burlew et al., 2000).

This not only has relevance for functioning in childhood but for longer-term functioning and social maturity. As previously mentioned, chronically ill young adults are found to lead more dependent lifestyles than healthy peers (e.g. Kokkonen et al., 1993; Kokkonen, 1995), and poor social maturation has been related to disease severity, and family distress and social problems in the family in childhood (Kokkonen, Kokkonen, and Moilanen, 2001). Chronically ill adolescents have also been found to be more likely than their healthy peers to be close to their mothers, to have overprotective parents, and to have parents that are very strict (McAnarney, 1985). They have also been found to be given little responsibility for household chores by their parents despite being able to carry out the task (Hayden, Davenport and Campbell., 1979), to be limited by parents in their involvement in school and social activities due to concerns about health (Wallander and Varni, 1998), be more externally controlled than well adolescents (Kellerman et al., 1980), and to feel domineered by the adults in their lives (Skar, 2003). Indeed, parents can often encourage dependency in their chronically ill children and fail to expect them become independent adults (McAnarney, 1985; Kellerman et al., 1980). However, while family and parenting factors can put chronically ill children at increased risk of

adjustment problems they can also serve as resistance factors, buffering the effects of illness-related and psychosocial sources of stress and equipping youngsters with essential coping resources. Intrapersonal factors like self-esteem and self efficacy form a vital component for adaptation in the face of chronic illness, and their development is determined throughout childhood and adolescence by crucial contextual factors.

### **2.2.2. Resistance Factors**

#### **2.2.2.1. Intrapersonal Resistance Factors**

In general terms, a number of dispositional variables have been associated with coping with stressful events. One such variable has been termed 'hardiness' and has been found to buffer the effects of stress on health (e.g. Kobasa, Maddi, and Kahn, 1982), having an internal locus of control has been found to have a similar effect on the effects of stress on psychological distress (Johnson and Sarason, 1979), and self confidence has been associated in health outcomes and on-going psychosocial functioning (Holahan and Moos, 1985, 1986). Further dispositional characteristics, such as an easy-going temperament, have also been suggested as resistance factors to stress. Individuals who describe themselves as easy-going have been found to adapt better to stressors than those who describe themselves as less easy-going (Holahan and Moos, 1985). They are also more likely to respond to stressful events without experiencing strain and to rely less on avoidance coping (Holahan and Moos, 1981, 1983), which has generally been associated with poorer individual adjustment in both children and adults (e.g. Holahan and Moos, 1987; Moos et al., 1990; Causey and Dubow, 1992). In the chronic illness literature in particular, a number of other intrapersonal factors have been discussed in relation to adaptation, and self-esteem and self-concept are two variables that have been particularly highlighted as having a significant impact on the adaptation of chronically ill children and adolescents (Anderson, Clarke and Spain, 1982; Thompson et al., 1993; Wallander et al., 1989; Burlew et al., 2000). Thus, the way in which chronically ill and disabled young people value and see themselves is seen as a key predictor of adjustment and success in later life (Kapp-Simon, 1986).

It is suggested that self-esteem affects stress processing and adaptation by influencing an individual's expectations (Thompson et al., 1993). For example, in young people affected by Spina Bifida, self-worth has been found to serve as a mediating variable for the effect of the physical appearance aspect of self-concept on depressed mood (Appleton et al., 1997), and higher levels of self-esteem and self-concept have been linked with overall adaptation (Moise, Drotar, Doershuk, and Stein, 1987; Burlew et al., 2000). Other intrapersonal variables such as social assertion and self-reliance have also been related to the psychosocial adjustment of chronically ill children and adolescents. For example, having an internal locus of control has been associated with adjustment to chronic illness (Krause and Stryker, 1984; Caldwell, Pearson, and Chin, 1987; Livneh et al., 2004), as has a sense of adaptive competence (Hurtig and White, 1986), and greater social assertiveness and higher levels of self-esteem have been associated with lower levels of anxiety and depression amongst chronically ill adolescents (Burlew et al., 2000). In terms of locus of control, it is posited that perceptions of control increase coping efforts and persistence, which reduces stress and affords the individual a more positive self image. This in turn has a positive affect on health (Bandura, 1977; Lefcourt, 1976; Thompson, 1981). Consequently, it is advocated that a significant amount of the variance in adaptation to chronic illness can be accounted for by a number of intrapersonal variables (Thompson et al., 1998).

In terms of the self-concept of chronically ill children and adolescents research has found mixed results. Adolescents with sickle cell disease have been found to have diminished self-concept as a result of their condition (Kumar et al., 1976) and greater dissatisfaction with body image (Morgan and Jackson, 1986). Studies of adolescents with chronic conditions such as cerebral palsy and spina bifida have also shown them to have lower self-concept than their healthy peers (e.g. Appleton et al., 1997; Shields, Murdoch, Loy, Dodd et al., 2006). However other research exploring spina bifida, cerebral palsy, and cleft lip/palate has reported no difference between scores on measures of self-concept, self-efficacy, and attitudes toward disability for adolescents with and without chronic illness or disability (e.g. King et al., 1993). Similarly, other research of adolescents affected by sickle cell disease has reported no difference in self-concept between the chronically ill adolescents and their healthy peers (Lemanek, Moore, Gresham, Williamson et al., 1986; Noll et al., 2006), adolescents with other chronic conditions (Kellerman et al., 1980), and national norms of diverse age



categories (Hurtig and White, 1986). More recent research has also shown chronically ill adolescents to not see themselves any less positively than healthy peers, and to not consider themselves to be different from other adolescents (Skar, 2003).

#### **2.2.2.2. Social-Ecological Resistance Factors**

##### **2.2.2.2.1. Parenting and Family Factors**

For over two decades family functioning has consistently been identified in the chronic illness literature as an important predictor of the psychological functioning of chronically ill children (Finney and Bonner, 1992; Quittner and Digirolamo, 1998; Wallander et al., 1989). Family factors in particular may serve to either exacerbate or attenuate the impact the disease has on the individual (Stuber, 1996; Ostroff, Ross, and Steinglass, 2000). In particular, parenting factors and family characteristics contribute significantly to the development of a high sense of self during adolescence, and intrinsic attributes of self-concept and coping style are invariably influenced by both familial and social factors. It is posited that ‘adolescents who are disabled may develop excellent self-esteem if they feel supported by their families and can take advantage of opportunities to develop their own identity and independence’ (McAnarney, 1985:100). This is reiterated by Antle (2004) who found parent social support to be a predictor of self-worth in young people with physical disabilities. Indeed, positive family characteristics have been found to contribute to children’s healthy adaptation whether they are affected by disability or not. These include parental self-esteem (Perrin, Ayoub and Willet, 1993), and family support (Holahan and Moos, 1987; Varni et al., 1991). Similarly, amongst children affected by chronic illness, family relations and the parent-child relationship have been positively related to healthy adjustment (e.g. Hurtig et al., 1989; Burlew et al., 2000), as has family support (Wallander and Varni, 1998), family cohesion (Kliewer and Lewis, 1995), and a family environment that is high in organisation and control (Burlew, Evans, and Oler, 1989).

Children in supportive, socially integrated, and well organised families have been found to be more socially competent and to have fewer behavioural problems (Moos

and Moos, 2002). When a child is affected by chronic illness or disability, a family environment that is supportive and organised is said to facilitate healthy adaptation, self-esteem, social competence, self-reliance, and psychosocial adjustment (Mink and Nihira, 1986; Moos and Moos, 2002). Independence and recreational orientation have been associated with better adolescent adaptation to living with chronic illness, especially in terms of social competence (Moos and Moos, 2002). Furthermore, social integration has been found to enhance child self-reliance (Mink and Nihira, 1986). Among other family characteristics, recreational orientation has also been linked with better school adjustment. High family cohesion, an orientation towards recreational pursuits, and high organisation have all been associated with self-reliance and competence amongst children with developmental disability (Nihira, Meyers and Mink, 1980). Family cohesion has also been linked to self-esteem and psychosocial adaptation in children affected by developmental disability (Mink, 1986).

Family integration; family support and esteem building; recreation orientation, control, and organisation; and optimism and mastery are four family recovery factors that have been identified as having a direct relationship to promoting successful developmental and physiological changes in children affected by cystic fibrosis (McCubbin, Patterson, McCubbin, Wilson, and Warwick, 1983; McCubbin, Thompson, Thompson, and McCubbin, 1993). Furthermore, for conditions that require complex treatment regimens, including cystic fibrosis, family factors have also been found to contribute to child and adolescent patients' compliance with treatments (e.g. DeLambo, Ievers-Landis, Drotar, and Quittner, 2004), which is important to the subsequent health of the child (Patterson et al., 1993) and to self-esteem, self-efficacy, and coping (e.g. Gold, Treadwell, Weissman, Vichinsky, 2008). Family functioning characteristics that have been related to adherence in this group of adolescents include higher family organisation and expressiveness, lower family attendance at social activities (Patterson, 1985; Patterson et al., 1993), and the ability to plan family activities around self care tasks (Ricker, Delamater, and Hsu, 1998). For children with diabetes fewer recreational activities and greater emotional expression is also related to compliance with treatments (Patterson, 1985), as are communication, problem-solving and conflict resolution skills (Bobrow, AvRuskin, and Siller, 1985; Wysocki, Miller, Greco, Harris et al., 1999; Hauser, Jacobson, Lavori, Wolfsdorf et al., 1990). Family functioning and family quality have also been

related to adherence in other paediatric conditions (LaGreca and Schuman, 1995; DeLambo et al., 2004).

#### **2.2.2.2.2. Social Support**

As well as family support peer relations are critical to the social and emotional functioning of children and young people, particularly peer acceptance and the support of close friends. Peer acceptance, which pertains to the degree to which children are accepted by their peer group, bestows children with a sense of companionship, intimacy, and self-esteem (Furman and Robbins, 1985). Even among healthy populations, peer relations can be a good indicator of current as well as long term emotional adjustment (Hartup, 1983; Parker and Asher, 1987). While much research of the social competence of chronically ill and disabled children has reported interpersonal difficulties and peer exclusion (e.g. Noll et al., 1991), other work has found no differences in their perceptions of peer acceptance and social relations to that of healthy peers (e.g. Frankel, 1996; Brown, Armstrong et al., 1993; Brown, Kaslow et al., 1993; Lemanek et al., 1994). While self-perceptions of peer acceptance and social competence are important variables to consider, little is known about the role that peer relations, notably close friendships, may play in chronically ill youngsters' adaptation (LaGreca, 1990; Nassau and Drotar, 1997; Schuman and LaGreca, 1999). For adolescents with chronic illness such as diabetes close friends have been found to provide a significant source of emotional support for their adjustment to the disease. They have also been found to provide practical support with some of the day-to-day aspects of disease management and to help chronically ill adolescents feel accepted despite their condition (LaGreca, Auslander, Greco, Spetter et al., 1995; LaGreca and Thompson, 1998). Peer support is seen as being complementary to family support and together they are proposed as representing 'a unique and important factor in psychological adjustment' (LaGreca and Bearman, 2000:149). High levels of social support from both family and peers has been related to better adjustment for chronically ill children than the support from just one of those sources (Wallander and Varni, 1989). Future research into the way in which peer relations can be affected by chronic illness and how potential psychosocial problems may be averted by social relations would be useful.

### **2.2.2.3. Stress Processing Resistance Factors**

Stress processing includes cognitive appraisal and coping methods as discussed. As with parents, chronically ill children make a series of judgements about potential sources of stress and their resources to cope with them, and the important role that appraisals play in children's psychological adaptation to chronic illness has been advocated by research (e.g. Wallander et al., 1989; Brown, Kaslow et al., 1993; Lewis and Kliwer, 1996; Thompson et al., 1998). For example, greater perceptions of illness-related stress has been related to higher levels of child maladaptation (Thompson et al., 1998), and increased perceptions of illness severity has been related to lower self image in chronically ill adolescents (Leung, Steinbeck, Morris, Kohn et al., 1997). Conversely, cognitions such as hope and having a positive outlook, effective problem-solving skills, an internal locus of control regarding illness outcomes, and lower perceived levels of functional limitation have also been associated with more favourable adaptation across a variety of chronic conditions (e.g. Helgeson, 1992; Lewis and Kliwer, 1996; Livneh et al., 2004). Furthermore, family stress processing and other resistance factors have been seen to function as stress processing factors relevant to children's adaptation (e.g. Burlew et al., 2000). For example, good communication skills have been found to lead to the use of social support as a coping strategy, and associated with reduced anxiety (Burlew et al., 2000), and hope has been found to enable individuals to appraise illness-related stressors as less threatening (Snyder, Harris, Anderson, Holleran et al., 1991; Lewis and Kliwer, 1996). Hope in turn has been related to active coping and to better physical and mental health (e.g. Holleran and Snyder, 1990; Sigmon and Snyder, 1990). Furthermore, increased knowledge regarding the illness, family relations, and family cohesion have been conceptualised by researchers as functioning as stress processing factors that are associated with more optimum adaptation (Nash, 1994; Telfair, 1994; Burlew et al., 2000). Thus, individual and family factors enable children and young people to view their illness in a more positive light and to process stressors related to it in a way that makes them more manageable, meaningful, and comprehensible to the individual.

The coping styles that chronically ill children and adolescents use have also been associated with adjustment, as it is posited that how a child copes determines the degree to which illness-related stressors will impact on adjustment (Wallander et al., 1989). As with parents of chronically ill children, active and avoidant coping strategies have been explored in relation to adjustment. Active coping has been related to a number of outcomes for the child, including less reliance on and use of the healthcare system (e.g. Gil, Williams, Thompson, and Kinney, 1991; Lewis and Klierer, 1996), lower levels of anxiety symptoms (Lewis and Klierer, 1996), and to more favourable adaptation in general (Livneh et al., 2004). Conversely, less adaptive coping strategies such as negative thinking, distraction coping, and reliance on avoidance coping have been related to greater use of the healthcare system and internalising and externalising behaviour symptoms, depression and anxiety symptoms, and difficulties with concentration (Gil, Abrams, Phillips, and Keefe, 1989; Gil et al., 1991; Thompson et al., 1993; Thompson, Gil, Keith et al., 1994; Lewis and Klierer, 1996). Further research has not found any relationship between either adaptive or avoidance coping and adjustment however, and it has been suggested that children and adolescents may use both types of coping interchangeably depending on the problem, which may be adaptive (Casey et al., 2000). Health control beliefs are also important to young people's coping. Realistic perceptions of control over health outcomes have been associated with adjustment to chronic illness (Helgeson, 1992). As children approach adolescence they become increasingly aware of their own agency and control over outcomes (Compas, Banez, Malcarne, and Worsham, 1991), and the way in which young people develop health control beliefs is very much influenced by health history and family background, particularly the way in which families organise around illness issues, the focus they put on the illness, and their attitude toward it (Lau, 1982).

Family factors have also been linked to children's use of particular coping strategies. For example, family cohesion has been positively associated with more active coping in chronically ill children (Klierer and Lewis, 1995), and greater family adaptability has been related to less reliance on disengagement coping (Sharpe, Brown, Thompson, and Eckman, 1994). Whether or not children adopt the same coping strategies as their parents however, remains unclear. Some research has found similarities (e.g. Gil et al., 1991; Band, 1990), while others have not (e.g. Klierer and

Lewis, 1995; Sharpe et al., 1994). However, family risk factors and parental coping strategies operate as salient contextual factors in children's lives and less adaptive forms of coping used by parents can have an adverse effect on children's functioning (Holahan and Moos, 1987). While no conclusions have been reached regarding a relationship between parent and child coping styles, it would be prudent to view coping from within the family systems framework and consider that the 'the coping efforts of individual family members may affect and be affected by the coping efforts of other family members in addressing a specific problem' (Compas, Worsham, and Ey, 1992:18) and that children develop their own coping repertoires through observational learning processes. As has been demonstrated, while disease parameters, illness-related and psychosocial factors present risks for adjustment, the familial and social context in which the chronically ill child develops substantially influences the development of internal resources, which shape the individual's view of themselves, their condition, and their ability to cope, and thus buffer the effects of illness-related risk factors on adjustment (Wallander et al., 1989). Indeed, even in healthy families, parental risk factors and family support account for a significant amount of variance in children's psychosocial development and physical health problems (Holahan and Moos, 1987).

### **2.3. Summary**

The disability stress-coping model as devised by Wallander and colleagues (e.g. 1989) proposes that psychosocial factors buffer the impact a stressor may have on adaptation and thus serve as potential protective mechanisms. The current body of research that has drawn on risk-resistance models has identified a number of psychosocial variables that contribute to the variability in adaptation. These include intrapersonal factors, such as self-esteem, mastery, control, and self reliance; social-ecological factors such as family functioning, social competency, and social support; and stress-processing factors such as beliefs and attitudes, illness perceptions and perceptions of stress, and coping mechanisms. These are relevant to both parents and children. A number of illness-related and psychosocial risk factors have also been identified, including functional care strain and functional dependence, the day-to-day management of treatment regimens and everyday family responsibilities for parents,

and pain, discomfort and physical limitations for children. Psychosocial risk factors also include limited development of social skills, limitations to independence, feelings of isolation and difference from peers, and limited perceived competence. For parents such risk factors can include the pain of watching one's child suffer and experience social difficulties and developmental delays, and engulfment in the care-giving role. The research discussed has helped in conceptualising how resistance factors can buffer the impact risk factors have on both family and child adaptation, and how child adaptation is greatly influenced by that of their parents and the functioning of the family as a unit.

In the case of MPS IH post-BMT a number of issues discussed here hold relevance. In terms of illness-related risk factors, there are a number that might impact on the child psychosocially, including progressive physical disability, physical and adaptive functional limitations and dependence, ongoing pain and painful orthopaedic surgery, and repeated hospitalisations. Being affected by such a disabling condition since birth can also have implications for psychosocial stresses including the risk of social marginalisation and peer rejection, social limitations, increased dependence on the family, the risk of developing a learned helplessness response to life's demands, and social immaturity. Depending on the way in which the child copes with the stresses associated with the condition, and on other intrapersonal and social-ecological factors that might buffer the effects of that stress, some children affected by MPS IH post-BMT may go on to develop psychosocial and emotional difficulties. Indeed, for the parents of children affected by this condition, the devastating experience of an MPS diagnosis, coping with the decision to undergo the high risk treatment of bone marrow transplant, then facing an uncertain future both in terms of their child's quality of life and length of life can have an enormous impact on adjustment and in turn on parenting and family functioning. These factors, of course, then have implications for child adaptation.

Whether or not psychosocial resistance factors do indeed buffer the relationship between biomedical risk factors and adaptation amongst children and young people affected by MPS IH post-BMT is not known. No systematic research of the psychosocial outcomes of bone marrow transplant for this patient group has been conducted to date. The need to examine this issue is therefore long overdue. By

exploring the psychosocial functioning of this patient group and how it is related to characteristics of the individual, the condition, family, and other social-ecological factors, a better understanding of the psychosocial issues that may arise from this condition may be reached, and factors that might moderate individual and family adaptation identified. This in turn will help to hone future psychosocial research into MPS and related diseases and to develop appropriate intervention strategies that may help individuals and their families adapt to living with MPS IH post-BMT.

## **2.4. Aims and Objectives of the Present Study**

The study had two aims. Firstly to explore the experiences of families affected by MPS IH post-BMT in an initial exploratory phase using qualitative methods. Secondly, in the second (quantitative) phase, to examine the moderating effects of resistance factors (e.g. intrapersonal, social-ecological, and stress processing factors) on the effects of psychosocial and illness-related risk factors or stressors on adjustment outcomes for both parents and their children affected by MPS IH post-BMT. The exact variables that constituted both risk and resistance factors were informed by the first exploratory phase of the study.

The objectives of the study were therefore four-fold:

1. To explore the experiences of parents of children and young people affected by MPS IH post-BMT, identifying the rewards and stresses associated with the experience and the factors that moderate the effects of stress on parent adjustment.
2. To examine the degree to which intrapersonal, social-ecological, and stress processing resistance factors moderate the effects of psychosocial and illness-related risk factors on parent adjustment.
3. To examine the degree to which intrapersonal, social-ecological, and stress processing factors moderate the effects of psychosocial and illness-related risk factors on patient participant psychosocial adjustment.
4. To assess whether spouses differentially experience parenting stress, coping, and perceptions of child care needs, and whether potential discrepancies in



scores or a shared view has implications for parent well-being, and in turn, child psychosocial development by serving as a moderating factor.

In relation to phase two of the study it was initially hypothesised that psychosocial resistance factors would account for more of the variability in both parent and patient adjustment than psychosocial and illness-related variables in a national sample

## **2.5. Methodology**

The study employed multiple methods within a mixed methodology. It was conducted in two phases. Phase one was exploratory and used a qualitative design. Phase two was informed by the findings of the first phase and employed quantitative methods. Figure 2-2 (p84) illustrates the mixed methods design employed. Mixed methods is an approach to enquiry which combines qualitative and quantitative methodology and integrates both qualitative and quantitative data to provide a unified understanding of a research problem (Creswell and Plano Clark, 2007). It is a growing trend in research methodology which has been referred to as a ‘movement’, and much has been written about mixed methods research providing more than quantitative or qualitative research can alone. It has proved to be particularly compelling in health research as health behaviours are multiply determined, with rich individual and contextual detail (e.g. O’Cathain, Murphy, and Nicholl, 2007).

Quantitative and qualitative methods both have their strengths and weaknesses. Qualitative research provides a richness of data from which meaning and context can be yielded (Creswell, 2008). It can be particularly useful in early research for understanding a complex phenomenon and for developing rapport with a particular community. Quantitative research on the other hand is less labour intensive, facilitates the collection of data from large samples, and across large geographical areas, and allows greater generalisability (Creswell, 2008). Mixed methods intelligently and strategically combine both methodologies and in doing so the strengths of both approaches are combined. This allows for a broader understanding of a research problem and for a more complete picture to be obtained (Creswell and

Garrett, 2008). It also produces findings that appeal to different audiences, as one or the other approach is not always appealing to all.

Mixed methods were used in the present study in response to qualitative and quantitative research questions, and a sequential exploratory design employed (Creswell et al., 2003). The study was therefore conducted in two distinct phases, and the first phase supportive and informative of the second phase, which was dominant. The study as a whole therefore fit within a positivist framework. The first phase employed qualitative methods in the form of semi-structured interviews, which were administered to parents and explored the experiences of families of children affected by MPS IH post-BMT. Interviews were tape-recorded and transcribed. The data yielded were subjected to Interpretative Phenomenological Analysis (see below and the next chapter for details). The emergent themes were used to inform the design of the second phase of the study, a survey that comprised a number of psychometric measures and demographic questions. Hierarchical multiple regression analyses were employed to measure the predictive value of a number of risk and resistance factors to parent and patient adjustment outcomes. A favourable opinion was received from the Eastern Multi-Research Ethics Committee, which granted ethical approval for this research on 2<sup>nd</sup> August 2004. REC reference number: 04/mre05/023

## **2.6. Recruitment**

Recruitment and sampling of participants was carried out in two phases. Eligible participants for both phases of the study were parents of children affected by MPS IH post-BMT. However, children and young people affected by the condition were also included in the second phase. Participants were recruited through the Society for Mucopolysaccharide Diseases (MPS Society), as they have an extensive database of MPS sufferers in the UK, one that includes the majority of families affected by MPS IH post-BMT. They work closely with all of the tertiary medical centres in the UK that diagnose, treat, and give ongoing care to individuals affected by the condition, and thus have an up to date record of all current cases. They are also in regular contact with many of the families, providing a quarterly newsletter, the opportunity for families to participate in activities, invitations to conferences and away-days, and the

provision of support and advocacy when required. Throughout the research close and regular contact was maintained with the five tertiary medical centres in the UK that specialise in lysosomal storage disorders and manage the care of all MPS IH patients in the UK. This was for the purpose of aiding recruitment through attendance at clinics and of making contact with difficult to reach families. As this area of study has been long neglected with this patient group, the medical professionals involved with individuals affected by MPS IH post-BMT and their families gave their full support and assistance where needed. MPS IH is after all a rare condition. There are thus a small number of individuals in the UK affected by it. All cases are therefore known to the five tertiary medical centres in the UK that specialise in lysosomal storage disorders and in turn to the MPS Society.

## **2.7. Analysis**

The analysis of this study was carried out in two distinct phases, reflecting the study methodology. In phase one, the interviews with parents were analysed using Interpretative Phenomenological Analysis (IPA) (Smith and Osborn, 2003). IPA is a method that stems from phenomenology, a philosophical approach that focuses on the subjective experiences of individuals and considers social, cultural and historical contexts in an individual's interpretation of the world. Phenomenology's concentration on subjective experience appeals to psychological research, as it allows for the investigation of the diversity and variability of human experience (Willig, 2008). Similarly to phenomenology, IPA aims to capture individual experiences. It however also recognises the role of the researcher within the research and analytic process and sees the analysis as a product of the interactions between the participants and the researcher (Smith and Osborn, 2003; Willig, 2008).

IPA is a well structured process that allows the in-depth examination of the specific phenomena under investigation. It is specifically designed to allow the investigator to draw from an interview the individual's perception of their particular situation and how they make sense of their personal and social world. As a tool of analysis it is also well suited to investigating novel areas (Smith and Osborn, 2003). Research employing this method of analysis often explores issues and events that are

particularly pertinent for the participants. Studies using IPA therefore tend to deal with major life altering or threatening events, conditions, or decisions (Smith, 2004). It is therefore considered to be a particularly appropriate method of analysis for use in this study. Furthermore, the use of qualitative methods has been called for by Wallander and Varni (1998) so that a better understanding of the experiences of having a chronically ill child may be reached.

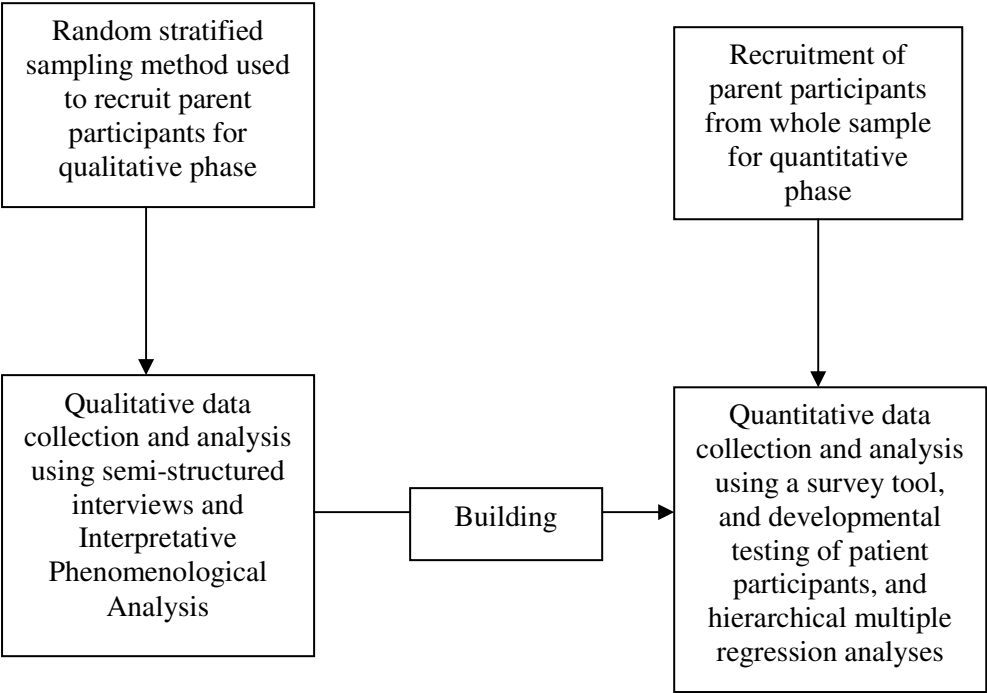
The super ordinate themes that emerged from the qualitative data informed the design of the second phase of the study, which employed quantitative methods. They were used to theory-build by identifying risk and resistance factors pertinent to parent and child adjustment to living with MPS IH post-BMT. A survey tool was created which comprised a number of psychometric measures and demographic questions. These were used to examine the experiences of both parents (mothers and fathers), where possible, and the children and young people affected by MPS IH post-BMT in a more structured and systematic way. Children and young people affected by the condition were also administered questionnaires and developmental tests in this second phase of the study. Data collected were analysed using hierarchical multiple regression analyses in order to examine the degree to which resistance factors moderate the effects of psychosocial and illness-related risk factors on parent and patient adjustment.

## **2.8. The Study's Contribution to Knowledge**

The experiences of families of children affected by MPS IH post-BMT have not been studied to date, and the psychosocial functioning of children and young people affected by this condition is a topic of study that has been neglected. Thus little is known about individuals' experiences and quality of life as they grow and develop through childhood into adulthood affected by this rare and severe condition. Furthermore, the disability stress-coping model has not been used to explore the experiences of this patient group and their families or to examine the factors that contribute to the affected children's psychosocial functioning. It has also been rarely applied in its entirety to evaluate the processes of adaptation to chronic illness. It is therefore a unique study on a number of levels and will serve to inform professionals

and researchers in the field of MPS and related diseases and the wider area of childhood chronic illness so that future research can be honed and progressed and appropriate support given to families.

**Figure 2-2. Flowchart for Sequential Exploratory Mixed Methods Design** (for a more detailed flowchart see Appendix A)



## **CHAPTER THREE**

### **QUALITATIVE PHASE**

#### **3.0. Methodology**

Phase one of the study was exploratory and therefore adopted a qualitative design. The rationale for this was that the psychosocial outcomes of BMT for individuals affected by MPS IH and the experiences of the parents who care for them are little understood. It is therefore a novel area of study, the exploration of which is well suited to qualitative methods (Smith and Osborn, 2003). The aim of this stage was to develop a better understanding of the rewards and challenges that parents experience in their daily lives and of the psychosocial issues that may arise from the condition for the individuals affected by it. Because the experience of having a child with MPS IH post-BMT can be emotionally taxing this was achieved through one-to-one audio-recorded semi-structured interviews with parents of children and young people affected by the condition. These interviews allowed the collection of in-depth information about parents' experiences from their child's diagnosis to the present day within a short period of time.

The data were analysed using Interpretative Phenomenological Analysis (IPA) (Smith and Osborn, 2003). As mentioned in the previous chapter, IPA is a method that stems from the philosophical approach of phenomenology, which focuses on individuals' subjective experiences of the world, and considers these experiences to be influenced by social, cultural, and historical contexts. It is an approach that is useful to psychological research as it allows for the investigation of the diversity and variability of human experience (Willig, 2008). IPA is also a well structured process that allows the in-depth examination of the specific phenomena under investigation. Although IPA shares the aims of phenomenology in terms of aiming to capture individual experiences, it also recognises the role that the researcher plays in the research and analytic process. From this perspective, the analysis is considered to be both phenomenological and interpretative as it is a product of the interactions between participants' accounts and the researcher's interpretations of participants' accounts. Thus, the analysis is dependent upon the analyst's meaningful interpretation of how participants make sense

of the world. Such interpretations are inevitably based on the researcher's own conceptions, beliefs, expectations and experiences (Smith and Osborn, 2003). IPA therefore requires reflexivity from the researcher who is expected to illuminate the analysis by presenting his or her own viewpoint (Willig, 2008).

When undertaking IPA there are some fundamental assumptions to consider regarding the individual and their world. Firstly, it is important to note that IPA considers what respondents say to hold some significance and 'reality' for them and represents a manifestation of their psychological world. IPA therefore assumes that narratives are related to beliefs or psychological constructs, though not necessarily directly or transparently related, and that meaningful interpretation of such conceptions can be achieved (Smith, Flowers, and Osborn, 1997). Secondly, experience is viewed as subjective since it is a product of cognition and perception, and different people have different perceptions of experiences. IPA recognises, however, that meanings are negotiated within a social context and that subjectivity can be affected by social interactions (symbolic interactionism). Finally, IPA functions from the premise of hermeneutics, suggesting that an understanding of the participants' experiences can be gained through the interpretations of the researcher (Smith and Osborn, 2003; Willig, 2008).

IPA enables predominant themes to be linked within and across cases. It is therefore an idiographic approach to analysis, which allows the researcher to discuss particulars building up to generalisations (Smith, Harre, and Van Langenhove, 1995). Moreover, IPA allows the discussion of the analysis in relation to existing literature, which facilitates the illumination of theories. IPA is also inductive, which allows for the emergence of unanticipated themes and responses. Being inductive is purported as being a central feature of IPA.

IPA was initially used in the field of health psychology (e.g. Osborn & Smith, 1998; Senior et al., 2002; Smith, 1996) in order to analyse qualitative data reflecting participants' experiences. It has also been used in social and counselling psychology research (Golsworthy & Coyle, 1999; Macran, Stiles, and Smith, 1999; Touroni & Coyle, 2002; Turner & Coyle, 2000). IPA is a particularly appropriate means of analysis for this research because it focuses on a previously unexplored condition and is

therefore a novel area of research. Adjustment to living with childhood chronic illness has also been demonstrated as being a subjective experience and subject to individual appraisals and coping resources.

### **3.1. Participants**

Eligible participants were those who were the main parent carer(s) of a child or young person with MPS IH post-BMT. All were members of the MPS Society in the UK. Eight interviews in total were carried out. In six of these an individual parent was interviewed, and in two of the interviews both mother and father were interviewed together. A total of ten participants were therefore interviewed. All names and identifying information have been removed. The majority of participants were female (8 out of 10). The affected children's ages ranged from 3 to 24 years. In line with the guiding principle for sample selection in phenomenological studies (Cresswell, 2007; Patton, 2002), the sample size was small, yet was purposefully selected to reflect the large age range of this patient population (currently 15 months to 24 years). While the purpose of this phase of the study was to gather extensive detail about each of the individuals studied and the generalisability of the findings not the concern, its purpose was also to gather information that can be used to inform the design of a survey tool suitable for research with a larger population of this patient group and their families. So that this tool may explore issues pertinent to a wide age range of patient participants, it was therefore imperative that the experiences of parents of children and young people at a variety of ages were explored. Age groups were therefore chosen as follows: 0-5 (young childhood); 6-11 (middle childhood); 12-18 (adolescence); 19+ (adulthood), and two families from within each age group randomly selected. A random stratified sampling method was therefore employed. A sample of eight families was interviewed as follows:

Group 1: Mother of 5 year-old male; Mother of 3 year-old female.

Group 2: Mother of 7 year-old male; Mother and Father of 11 year-old female.

Group 3: Mother and Father of 12 year-old male; Mother of 16 year-old female.

Group 4: Mother of 24 year-old female; Mother of 24 year-old male.



### **3.2. Data Collection**

Parents were invited to volunteer to participate in the study by letter (see Appendix B). They were provided with an Information Sheet (see Appendix C), which explained the aims and procedure of the study. They were informed that the study was to comprise one interview, which would take no longer than two hours and would take place in their home at a time of their convenience. They were informed of the nature of the interview questions, and that the interview would be tape-recorded and subsequently transcribed. They were assured of anonymity, and that they could withdraw from the study at any time. Seven of the eight reply-slips were promptly completed and returned to the researcher. One family did not respond and were removed from the study. An alternative family was randomly selected from the MPS Society database, from within the relevant age strata, and were approached using the method described above. This family duly volunteered to take part in the study. Once the completed reply-slips had been received, the potential research participants were contacted by telephone and an appointment made for the researcher to visit their homes. Potential participants were given the opportunity to ask any questions they wished about the research. A contact telephone number for the researcher was given to each family.

All interviews took place in the participant's home and lasted approximately two hours. All interviews were conducted by the author and were tape-recorded. The interviews began with an opportunity to read the Information Sheet again and to ask any questions about the study. Participants were then asked to sign a Written Informed Consent form. The Written Informed Consent Form (See Appendix D) stated that participants had read and understood the aims and procedure of the study and that they were willing to take part. It also stated that participants gave permission for their child's GP and bone marrow consultant to be informed of their participation in the research (See Appendices E and F for GP and Consultant letters). Participants were reminded that they could withdraw from the study at any time and that they were not obliged to answer any questions with which they were not comfortable. Participants were not paid to take part in this phase of the study.

An interview schedule was designed. Its design was informed by the author's personal communication with MPS families and with medical professionals and family support group workers who work closely and extensively with this patient group, their carers, and families. It was also informed by previous research into the psychosocial sequelae of living with childhood chronic illness. The interviews were semi-structured and consisted of a series of open-ended questions, with most followed by subsidiary prompts. This type of interview is recommended for qualitative research as it allows for the researcher and participant to engage in a dialogue, and for the researcher to be flexible in their questioning and use of prompts in response to the participants responses (Smith, 1995). The questions covered a variety of areas including the family's experience of the MPS diagnosis, BMT decision-making, expectations of BMT, daily stress and coping, social support, parent's perceptions of the child's disability, parent's coping self-efficacy, and parent's perceptions of the effects the illness and treatment experience has had on the family. Parents were also asked their thoughts on their children's psychosocial well-being, including whether or not they felt they had any behavioural, social, or psychological difficulties. The questionnaires were piloted with the first two families interviewed then, since they appeared to work well and no changes deemed necessary, the data from these interviews incorporated in the main phase one study.

No participants withdrew from the study and none refused to answer any of the questions asked. Eight full interviews were therefore completed. No personal information was attached to the tapes. Tape-recorded interviews were transcribed and any references made to individual names were rendered anonymous. The transcribed interviews were coded and all coding information has been kept separately from the transcripts under lock and key.

### **3.0. Analytic Strategy**

The analysis of the transcripts involved a number of stages as suggested by Smith and Osborn (2003). These were as follows:

1. The first stage of analysis consisted of several readings of the first transcript. Notes were taken about key phrases felt to be of interest and significance, preliminary interpretations and any connections. Throughout the transcript, summaries of the meaning of what was being said were made and noted in the left hand margin.
2. The second analytic stage involved the transformation of these preliminary comments and summaries into emergent themes. These encapsulated the essential meaning of what appeared in the text and therefore of the participant's experiences. It required a higher level of abstraction and evoked more psychological terminology.
3. The third stage involved the compilation of a list of themes and an examination of the connections between them. Themes connected were grouped and given a descriptive label (super ordinate theme) and a table of themes compiled. As super ordinate themes were being categorised the source material was repeatedly check to ensure the super ordinate themes were consistent with the participant's account.

This process was repeated for the rest of the interviews. The super ordinate themes for each interview were then compared with one another and a table of initial comparative themes produced. For the most part themes were comparable across interviews. However, those that were not supported well by the data were dropped. The next stage involved the compilation of a master table of comparative themes, which formed the basis for writing up the results. The thematic content of the table was transformed into a narrative report of the participants' experiences with extracts from the text used to illustrate the interpretation.

### **3.2. Research Process and Rigour**

Some of the themes that were elicited from the transcripts followed the questions on the interview schedule, including those that relate to expectations of BMT, stress, support, and coping. However, others emerged repeatedly throughout the transcripts and related to a variety of areas. Examples of this are uncertainty, perceptions of child vulnerability, and the way in which parents used positive reframing to appraise their

child's condition and their situation. Since I presented myself to the families as a researcher employed by the MPS Society, a patient group that is dedicated to the support and care of individuals affected by MPS and related diseases, their families and carers, it is possible that some of the parents wished to convey a positive attitude regarding their situation and their child. This was not the case for all however, and I felt that parents felt very relaxed in my company and able to speak openly. I have been conducting research with families for many years and pride myself on being able to develop a rapport with individuals and put them at ease in a short space of time. However, prior to this particular piece of research beginning I attended a conference on MPS diseases, and other events, and attended BMT clinics at Manchester Children's Hospital, which families attended. I therefore met some of the families that participated in this first phase of the study before interviewing them. Parents therefore felt comfortable and familiar with me, and trusted that I had their best interests at heart as I was employed by the MPS Society to conduct the research. It is likely that the positive response for participation in the research was also due to these reasons. Furthermore, the psychosocial outcomes of BMT for this patient group is an area of study that has been neglected by researchers, but one that is of great interest to parents of children affected by this condition. Parents were therefore keen to be involved and to further our understanding of the challenges that individuals face growing up with MPS IH post-BMT.

Despite the interview schedule being quite prescriptive the interviews flowed very well and often exceeded two hours, with parents talking at length about their experiences. Although children affected by this condition receive a lot of medical attention, little attention has been given to family members regarding their feelings and experiences, or to the psychological aspects of living with this condition for the patient. Parents therefore relished the opportunity to talk about their feelings, with some expressing anger and frustration at the lack of support and understanding of others, and others expressing sadness and fear regarding their child's limitations, ill health, and their potential loss. However, as mentioned above, parents were for the most part extremely positive about their situation, in particular the health and well-being of their child, their survival and presence in the world. Even though they talked about a great deal of stress and distress associated with the experience of having a chronically ill child, they all

demonstrated enormous love and care towards that child and gratitude for their survival and relative good health.

With regard to validity and reliability the exercise is considered to be no less important in qualitative research as in any other form of research. However, the criteria for the evaluation of qualitative studies, as compared with the criteria for evaluating quantitative studies (i.e. replicability, reliability, large samples etc.), is different (Smith et al., 1995a, b). Indeed, researchers adopting the IPA approach in particular recognise the subjectivity of their analysis, as their main objective is to offer their own interpretations of participants' accounts. Objective reality it is argued cannot be applied to subjective experiences and accounts (Yardley, 2000). This notwithstanding, two important criteria suggested to assess the internal validity and reliability of qualitative research, are internal coherence and the presentation of evidence (Smith, 1996). Internal coherence relates to the question of whether the argument presented in the study is internally consistent and justified by the data. Evidence refers to verbatim examples from the interview transcripts which it is suggested should be presented in the paper to allow the reader to question the interpretation. As a check on my analysis, rigorous discussions took place with colleagues and my supervisors about the analysis process and strategy employed. Through this I ensured that the themes and my interpretations were supported by the data. The analysis section below also includes numerous extracts from the transcripts which further allow readers to assess the consistency between the data and my interpretation. In these quotations, material in square brackets is included for clarification purposes. The names of the participants, the people they refer to and location names have been changed to preserve confidentiality and anonymity.

### **3.3. Findings**

A number of super ordinate themes are reported here and shown in Table 3-1 overleaf. A full list of super ordinate and sub-themes can be found in Appendix G, and details of sub-themes are presented throughout the analysis under each super ordinate theme heading. The themes describe the impact that MPS IH has had on the child and the family from diagnosis in the child's infancy, through the bone marrow transplant also

in the child's infancy, the experience of the child's medical regimen from significant surgical procedures and hospitalisations to routine medical checks and monitoring, to issues relating to parenting, coping with day-to-day stresses, and support, both helpful and unhelpful. They describe parent's feelings of uncertainty in a number of areas, how they feel protective of their child, and their appraisal of their child's condition, the family's experience, and the situation they are in. They also describe parent's perceptions of their child's psychosocial and adaptive difficulties, and their children's understanding of the condition by which they are affected. Themes were based on the frequency with which issues were raised by parents and on the importance parents ascribed to them. Verbatim examples of participants accounts were selected as they best illustrated the themes identified. They are depicted in the table and in the following section in the order in which they appeared in the transcripts.

**Table 3-1. Master Table of Super Ordinate Themes**

1. Shock and disbelief at diagnosis
2. No decision to be made about whether or not to undergo BMT
3. Uncertainty
4. Having low expectations of the child
5. Adaptation as an ongoing process
6. Cognitive appraisal
7. Stress and coping
8. Social support
9. Lack of understanding
10. Wanting to protect the child
11. Parents' perceptions of child psychosocial well-being and development

## 1. SHOCK AND DISBELIEF AT DIAGNOSIS

<b>Sub-themes</b>	Comprehending the deterioration and death of a seemingly healthy child
	Difficulty digesting medical information at an emotionally devastating time

A number of parents interviewed talked about the shock of receiving such a devastating diagnosis for their child and reported how difficult it was to conceive of their seemingly healthy child deteriorating so dramatically over the coming years. This was clearly a devastating experience. They also described how difficult it was to take in and comprehend complex medical information, which was necessary for them to make an informed choice regarding treatment, at such an emotionally devastating time. These experiences have been described by previous research. Firstly, a sense of ambiguous loss and disenfranchised grief has been reported as being experienced by families when a family member is diagnosed with a genetic condition but is asymptomatic (e.g. Sobel and Cowen, 2003). This can have implications for coping and is an area of concern that requires attention from medical professionals and support organisations so that families can be appropriately supported at this time. In relation to the comprehension of complex medical information, the work of Jedlicka-Kohler and colleagues (1996) has described the experiences of parents of children affected by cystic fibrosis and how their ability to comprehend and remember medical information presented was impeded by the shock reaction to the diagnosis. This has implications for parents' understanding of the potential outcomes of BMT for their children affected by MPS IH in the longer-term and is again an issue that needs to be recognised by medical professionals to enable them to provide appropriate support to families. The mother of the five year-old male describes the diagnosis experience:

‘You just can’t begin to explain it the bottom just drops out of [your world]’

The mother of the seven year-old male similarly describes the shock experienced:

“It took the consultants until he was 10 months old to diagnose him. They sent us to [a specialist hospital] and hit us with a double-decker bus – he’s got MPS.”

As does the mother of the twenty-four year-old female:

“.....it was just complete devastation, but at the time you’re on such a roll of ‘this can’t be happening, there’s got to be a cure somewhere!’”

### **1a. Difficulty comprehending the deterioration and death of a seemingly healthy child**

Comprehending the grave and degenerative nature of the condition, when their child was born a seemingly healthy baby, was very difficult for parents. They talked about the difficulty they experienced understanding that their apparently healthy infant would follow the devastating natural course of MPS IH being described to them by medical professionals and that they would not live much beyond ten years of age. The mother of the seven year-old male describes this experience:

“We were quite dumbstruck, because all I remember him saying was ‘this is very, very bad’, and I’m sitting watching [my son] who was rolling about on the floor, perfectly fine by what I thought.”

The mother of the twelve year-old male talks of her disbelief at the information she was being given:

“My personal thoughts when we were told was utter disbelief. I just looked at [my son] and I couldn’t believe what the doctor was telling us.”

Similarly, the mother of the twenty-four year-old male explains how what she was being told did not make sense to her:



“It didn’t mean anything, two and two didn’t add, it didn’t mean anything to me.”

### **1b. Difficulty digesting medical information at an emotionally devastating time**

The mother of the five year-old male also explains how difficult it was to digest the complex medical information at such an emotionally devastating time:

“That’s the trouble. At the time when you need the most information it’s the time when it’s very difficult to take it all in. It’s very upsetting, all the details about clawed hands and feet, you can’t picture what your child’s going to be like and look like, and how they’re going to relate to you. No matter how much information you get, you can’t fully picture it.”

## **2. NO DECISION TO BE MADE ABOUT WHETHER OR NOT TO UNDERGO BMT**

<b>Sub-themes</b>	No expectations of BMT: Focus on saving the life of the child
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Despite parents’ little understanding about the natural course of the condition, being given scant information about the outcomes of the bone marrow transplant, and the high-risk nature of the treatment, which was the only viable treatment available to them, they were required to make a swift decision about whether or not they wished their child to undergo BMT. Most parents explained that there was no decision to be made. It was the only chance of survival for their child. Therefore, despite the risks, they felt they had no choice if they were to save the life of their child. For example, the mother of the seven year-old male explains how with such a poor prognosis she felt it was worth risking her son’s life in order to try to save it:

“I didn’t find it difficult at all, and I don’t think [my partner] did either, because our view was if we do get a donor, then that would be the decision made. [The Consultant] also explained that there was a high

risk of losing [our son] through transplant, but my view was that he was going to die anyway, and I would rather he died with me trying to help him than die with nothing. So that was how we came to the decision we did.”

Similarly, the mother of the twelve year-old male explains that there was no decision to be made if they were to try to save their son’s life:

“I don’t think there was ever any question for us, because we just thought if there’s a way of saving his life we’ve got to do it.”

This was again the case for the mother of the sixteen year-old female:

“There wasn’t a decision to make, it was just like, well that’s the only thing that’s going to help her, and we’re going to have to go with it. And when they say that it’s obviously a dangerous procedure and that she could die in the process, your options are that she either dies trying to make her better or she’s going to die within a few months anyway, so there’s no option.”

And for the mother of the twenty-four year-old female:

“I think that all that we were told was that there were no guarantees, and I think we talked to the family about it and really we came to the conclusion that there wasn’t a choice to be made, we were going to lose her anyway. [My husband] was like ‘oh we’ve got to go for it, we’ve got to go for it!’.”

Despite uncertainties regarding the outcomes of the treatment, families reported that the decision to allow their child to undergo BMT was made for them in the sense that they felt it was their only chance to save their child’s life. This reflects the view of Andrykowski (1994) who posits that the decision to go ahead with bone marrow transplant is made as soon as it is offered as a treatment option, particularly when the condition the individual is affected by is life-threatening. This is reinforced by the

following sub-theme and selection of extracts which illustrate how the will to save the life of their child over-rode any expectations parents had of the treatment.

### **2a. No expectations of BMT: Focus on saving the life of the child**

Mostly parents explained that they had no expectations of the BMT and that they did not think any further than the present and the task in hand, which was to ensure the survival of their child through the BMT. For example the mother of the seven year-old male says:

“I didn’t have a clue [what to expect from the BMT], I really did not have a clue. We’d never met another MPS child, we’d never seen any. I didn’t have any idea what we were in for or what he would be like afterwards. I was just ‘we have to do this or he’s going to die’, and that was it.”

The father of the eleven year-old says:

“We didn’t [have any expectations of the treatment]. The whole focus was on getting our little baby through the bone marrow transplant so that she’d live longer than six [years]. No expectations at all.”

Similarly, the mother of the sixteen year-old female states:

“All that you focus on is that your child lives through it. You don’t think that far into the future. At that position, when they are having the transplant, you just want them out of hospital and to be better, and you don’t think any further than that.”

### 3. UNCERTAINTY

<b>Sub-themes</b>	Uncertainty about outcomes of the BMT
	Uncertainty about making decisions on behalf of the child
	Uncertainty about the child's future psychosocial quality of life and adaptive functioning
	Uncertainty about the child's future well-being after the parents' death
	Uncertainty about allowing reasonable independence
	Uncertainty about future adult independence and issues relating to adult living

Parents' feelings of uncertainty were highlighted in a number of areas, including with regard to the outcomes of the BMT, outcomes of surgical procedures, their child's future psychosocial quality of life, the child's well-being after the parent's death, whether or not to encourage independence, and future adult independence and issues relating to adult living. Uncertainty has been highlighted by previous authors as having significant implications for parents' coping abilities as it is suggested to impact upon their perceptions and hinder the psychological management of a condition (Swallow and Jacoby, 2001). The diagnosis of a chronic condition has been found to present parents with a number of uncertainties, particularly when treatment options are high-risk, treatment outcomes uncertain, and future quality of life unpredictable (e.g. Cohen, 1997). Expectations about treatment and prognosis can therefore be greatly influenced by uncertainty (Hilton, 1992), and can have serious emotional consequences for parents of children diagnosed with chronic illness (Bonner et al., 2006; Gibson, 1995). Previous research has highlighted how parents can experience uncertainty in both acute and long-term contexts. This can be when making decisions regarding treatment, which constitutes acute uncertainty, and in relation to longer-term concerns such as those about the child's future well-being (Mishel, 1981; Bonner et al., 2006). These types of uncertainty are reflected in the extracts presented below, as are a number of emotional responses which have been associated with uncertainty, in particular guilt and worry. This is a pertinent finding as guilt and worry in particular have been positively related

to perceived impact on the family and parenting worries, and such emotions have been related to parents' perceptions of child vulnerability (Bonner et al., 2006).

### **3a. Uncertainty about the outcomes of the BMT**

As stated above, uncertainty was a super ordinate theme that emerged in many different areas of the texts and related to different aspects of the families' experiences. Initially however, parents spoke of their uncertainty regarding the outcomes of the bone marrow transplant and how medical professionals could give no guarantees or certainties regarding outcomes. The mother of the seven year-old describes the limited knowledge she has about the longer-term outcomes of BMT:

“There wasn’t very much they could tell us because there wasn’t a lot of children that had been transplanted, they were still quite young. The main things were that they would have major surgery on their bones and joints.”

Similarly, the father of the twelve year-old male explains how the medical professionals are unable to give firm prognoses:

“Nobody will give you a prognosis, nobody will say really, because they all say that every child is different.”

The mother of the sixteen year-old female expands on this and describes how she appreciated that the medical professionals did not have enough experience of MPS IH patients undergoing BMT at the time to give information about outcomes with any certainty:

“It was very much ‘don’t know’, everything was ‘don’t know’. ‘We don’t quite know what the outcome is going to be’, ‘we don’t know how it’s going to affect her’, which is fair enough because they didn’t, they hadn’t had many children go through it”.

The mother of the twenty-four year old female spoke about the uncertainty experienced at the time of the diagnosis and treatment and wonders on reflection whether with more certainty about the outcomes of the BMT the strain of caring for their daughter would have, or could have, been any different with more information:

“I suppose I think ‘why did life have to be so hard, so difficult to cope with?’ and I think maybe it’s nice to know what you are letting yourself in for, but could it have been any different? Did the doctors keep anything from us? I don’t think really they did, because they didn’t know.”

### **3b. Uncertainty about making decisions on behalf of the child**

Having made the difficult decision to go ahead with the BMT, families were then faced with the prospect of making further difficult decisions for their child. In particular, since skeletal abnormalities continue to progress despite successful engraftment of the BMT, orthopaedic surgical procedures can be considered to correct such abnormalities and potentially prolong mobility. However, these procedures do not have guaranteed outcomes, particularly for this patient group, as the skeletal problems are progressive and may reoccur. There are also no guarantees that the surgery will be of benefit to the patient. Families therefore face a difficult dilemma and expressed guilt and uncertainty when faced with making such significant decisions for their child. The mother of the five year male old describes this:

“My fears for the future are all the orthopaedic side, and again, because it’s ‘guinea pig’ time and obviously the doctors are quite open about the fact for them it’s new and they don’t know, so again I’m very fearful of the decisions I’ve got make on his behalf without much knowledge.”

She goes on to say:

“We don’t know if we’re going to put him through stuff that’s meaningless or put him through stuff that will be beneficial, and he’s the one that’s got to go through it. So that’s going to be very difficult.

Will he in ten years time say, 'I didn't want that operation thank you that hurt loads and I'd rather be in a wheelchair but without the pain'. Will he be in a wheelchair and say 'why didn't you do my knees? I love running around I wish I was still active'. You could do all of it and he still doesn't end up active, you just don't know, so that is horrendous the thought of that."

The mother of the seven year-old male explains how as her son gets older she hopes that she can involve him in the decision-making, which could serve as a guard against guilt. Indeed, she describes how difficult it was to put her child through such a traumatic experience and how guilty she felt having to take the responsibility for making such decisions:

"I don't know what it'll be like with the next lot of surgery if he gets any. I'm hoping that I can discuss it with him, and he can hopefully then make some of the decisions. He's not old enough to say, 'I don't want that done' or 'yeah I can' but hopefully we can discuss it and he'll understand it now, and be a bit more comfortable with things. Before this it's been 'well you took me hospital and you let all these people hurt me and it's your fault' which I found really, really hard, and for [my son] to be that upset it's awful, awful."

### **3c. Uncertainty about the child's future psychosocial quality of life and adaptive functioning**

Many parents expressed concern about their children's uncertain future psychosocial quality of life. With the knowledge that their children's physical disabilities would progress and learning disabilities become more apparent, many parents expressed concern about their child's social world changing and becoming more challenging for them. For example, the mother of the five year-old male expresses concern that as her child gets older he will get left behind by his peers and become isolated:

"The thing that worries me for the future, it's all very well now, he's got this set community and it obviously comes through me and [my

husband] as well and my friends, and I think there'll be this time where his peers outgrow him but don't fully understand his difficulties."

The mother of the seven year-old male expresses similar concerns:

"My only concern is that by that time you will see the disabilities, and I hope he's still got a good circle of friends, like he's got now, and I hope they don't dwindle away as his disabilities become more apparent. That's my only concern really."

The mother of the eleven year-old female describes her uncertainty about her daughter's future cognitive and functional development and her ability to cope throughout the education system:

"I can see problems [in the future] and worry about them now, and then the stress level goes up again. I think the main thing at the moment is her education, if she's ever going to be able to read and write, and what it's going to be like for her when she gets to secondary school, that's the main one at the moment."

### **3d. Uncertainty about the child's future well-being after the parent's death**

Parents also described their uncertainty about what would happen to their child if they were to pre-decease them. They expressed concern about how they would cope and how they would be cared for. This was particularly a cause for concern for the parents of the older children and young adults. The mother of the 12 year-old male explains:

"A lot of worry about the future obviously, because he's so special to us, we don't want to lose him. Also I think one of my worries is... how things will be when we're not around. I've got to learn to take one day at a time, I'm trying to do that, but sometimes I do worry long-term about the future and how it will be for him."



The mother of the 24 year-old female draws attention to her fear that her daughter may be institutionalised after her death:

“I do hope that I survive [my daughter].....it doesn’t worry me, it’s not something that I worry about all the time, I haven’t got any control over it, but I hate to think of her being in an institution....”

The mother of the twenty four year-old male expresses similar concerns and hopes that they have prepared for this eventuality adequately:

“My biggest thing is that I hope I outlive him, for his sake, not for mine obviously. If I don’t I hope we’ve put enough things into gear to cope with it.”

### **3e. Uncertainty about allowing reasonable independence**

The issue of independence was talked about by most of the parents. Many parents expressed uncertainty regarding their child’s level of functioning and considered aspects of independence as being subject to this as a condition. The mother of the youngest child, the three year-old female, expressed her willingness to allow her daughter to go out and socialise when she gets older. She however found it difficult to conceive of her child as being socially mature enough. She expresses some uncertainty regarding this and sets the condition of social maturity on her ability to conceive of her daughter’s future independence:

“Yeah I’ll be fine, if she’s sociable enough, mature enough. Yeah I’d be fine with that. I’d let her go out and have her freedom and whatever she wanted to do. Well not whatever she wanted to do, but I’d let her go.”

The mother of the eleven year-old draws attention to the fact that her daughter has been limited by the protection of her family in the development of her independence. She expresses uncertainty regarding her competencies and whether she would be safe outside of the home without supervision:

“I worry more about things. An easy example is her riding a bike, another example is crossing the road. She’s been brought up in a way that she hasn’t learnt the independence that other children have.”

Similarly, the mother of the twelve year-old boy expresses her uncertainty regarding her son’s safety outside of the home without supervision:

“But I just feel that I wouldn’t be able to let him out on his own at the moment, to cross that road to go to the shop. I’ve walked behind him to see if he does it, and he is very good, but....”

### **3f. Uncertainty regarding future adult independence and issues relating to adult living**

When asked about their expectations of their children’s future adult lives, including the issue of future independence from the family, the majority of parents expressed uncertainty. The mother of the three year-old female describes how she would like her daughter to have a job, but is realistic about the likelihood that she will continue to require support and has lower expectations of the type of job she might acquire:

“Well, she’s still going to need that extra support, so I don’t know how she’s going to [get on]. I’d like her to eventually go out and get a job, a job that she’s able to cope with.”

The mother of the five year-old male expresses her wish that her son experiences an adulthood that is independent from the family, but also describes her uncertainty regarding whether or not her son will ever leave home and how much support he would require if he did:

“You hope that he would end up as an independent adult, whether that would be living with others, with support, or whether he would.... I would like him to be independent, but he may be with us forever, I don’t know, but I would strive to that.”

The mother of the 12 year-old male, like the mother of the three year-old female, can conceive of her son living independently from the family, but sets his maturity and mobility as conditions of this. She is also unsure of the type of accommodation he would be suited to:

“...looking ahead, as long as he’s different from what he is now, where he’s maturer and he’s able to get around, yeah I think I’d have no problem with [him living an independent life], I’d be thrilled in fact and it’s what I hope for in the future. I hope that he will be able to look after himself, but it just depends on whether it’s something that he can go off and do on his own, or whether he would have people with him, I just don’t know.”

Feelings of uncertainty about the outcomes of the BMT in the longer term and not knowing about the likely outcomes of disease management options such as orthopaedic surgery has serious consequences for the parents interviewed in terms of stress and was a particularly difficult problem for them to cope with. The fact that parents do not know whether or not their children will have a shortened life or how they are likely to function in adulthood physically, socially, emotionally, and adaptively created uncertainty and subsequent negative consequences for parents in terms of their coping abilities. It also has implications for perceptions of child vulnerability and parenting as previously discussed and as highlighted in the literature (e.g. Anderson and Coyne, 1991; Holmbeck et al., 2002). The extracts presented in the following section illustrate this. Without firmer knowledge of outcomes it is decidedly difficult for parents to parent with realistic long-term goals and expectations in mind and to prepare their children for adulthood, equipping them with the appropriate social and self-help skills. Though not necessarily the discovery of a new concern for parents of chronically ill children (e.g. Ievers and Drotar, 1996), these findings highlight the implications that the lack of knowledge about a rare condition can have for families. Again, it is important that medical professionals are made aware of these implications so that families can be appropriately supported in this area.

#### 4. HAVING LOW EXPECTATIONS OF THE CHILD

Previous research has highlighted the relationship between uncertainty and perceptions of child vulnerability (Bonner et al., 2006), and has illustrated how the employment of an over-protective parenting style can hinder the child's development of autonomous behaviour and inhibit the development of his or her future social maturity (e.g. Anderson and Coyne, 1991; Holmbeck et al., 2002; Kokkonen et al., 1993). The following extracts illustrate how some of the parents interviewed in the present study have low expectations of their child and parent them in such a way that they are hindered in their development of autonomous behaviour. The mother of the eleven year-old female points out exactly this and states that as a result of her parenting her daughter would not be equipped to live independently.

“I am in a lot of ways more overprotective over [my daughter] because I don't think that [my daughter] has that ability that [my younger daughter] has already gained. I can't see her ever reaching that standard too. Whether she can live independently I doubt very much.”

She goes on to describe her daughter's aspirations but does so with a tone of doubt and incredulity as if to say 'this is what she wants but it's not going to happen':

“She says to me she's going to work, she's going to be a vet..... We've got a flat here and she has said that that is where she wants to live, she wants to live in the flat.”

The mother of the sixteen year-old female explains how she feels her daughter would endanger herself if she lived away from home and expresses her doubts about this happening:

“I don't think she will [leave home] to be honest, I don't think she will. For one she couldn't cope, just simple things, even getting herself dressed. I think she would be a danger to herself the amount of things that she's done here. Just trying things with somebody looking on.

She's turned the gas cooker on before now.... So yeah, I can't see it happening myself."

The mother of the sixteen year-old also spoke of her uncertainty regarding her daughter's likelihood of meeting a romantic partner:

"I'd like to think that maybe she would [meet a partner] one day, but I can't see it, it's just not one of those things I think will happen. It's like one of those things you think 'oh it would be nice' but you know, I don't think it would happen. It's probably one of those little dreamy things up here somewhere."

## 5. ADAPTATION AS AN ONGOING PROCESS

<b>Sub-Themes</b>	Coming to terms with the diagnosis
	Acceptance of and adaptation to the child's limitations and aspects of the condition
	Balancing the care of the ill child with the parenting of well siblings

Evidently families found the experience of adapting to their child's condition challenging. It was described as being an ongoing process of ups and downs, where feelings of acceptance, control, and competency are challenged by new experiences that need adjusting to. The idea that coping is a non-linear process has been previously written about, and it has been described as a series of ups and downs where unexpected challenges present themselves and require recovery from and adaptation to (e.g. Spinetta, 1981; Wortman and Silver, 1987). The results from the present study highlight the difficulties parents experience coming to terms with their child's MPS diagnosis, their limitations, the commitment to care, and the likelihood that their child may not reach the goals parents might have expected of a healthy child. They also illuminate the issue of adaptation as a family process, illustrating how adjustments to the change in family dynamics need to be made by all family members, and how balancing the needs of sick and well children can pose a challenge. How families respond to having a child with chronic illness has been widely reported to influence both child and individual

family member's adjustment (e.g. Wallander and Varni, 1998). Equally, how the family functions as a whole has been related to both child and parent adjustment (e.g. Warfield et al., 1999).

### **5a. Coming to terms with the diagnosis**

All the parents spoke of the difficulty they experienced coming to terms with their children's illness and functional impairments. The mother of the twenty-four year-old female explains how her husband found it difficult to accept their daughter's diagnosis at first:

"I think my husband found it very difficult to accept. ....His immediate reaction was 'there's got to be a cure somewhere', that was it he wasn't going to accept it."

The mother of the twelve year-old male describes how it has taken a long time to accept her son's illness and limitations:

"I used to be a bit 'why has this happened? I want him to be ok', but I'm slowly changing the way that I think. I'm accepting things more."

The mother of the five year-old male explains how adjustment is an ongoing process that has its ups and downs:

"I think when your child's diagnosed with any sort of disability it takes you a long time. You slowly come to terms with it. You think you have [come to terms with it] and then something else happens and you [realise you] haven't quite, and as a family you very slowly change and come to terms with things."

## **5b. Acceptance of and adaptation to the child's limitations and aspects of the condition**

Post-transplant the outcomes vary widely and some children affected by MPS IH require more care than others. Some may experience medical difficulties, some learning disabilities, and others particularly severe physical disability. Many parents spoke of their adaptation to these differing aspects of their child's illness and how at times it has been difficult for them to come to terms with. For the mother of the three year-old female, the difficult aspect of her child's condition was the extra care needed (e.g. tube-feeding). In the short term following transplant some children affected by this condition can experience eating difficulties and require feeding straight into the stomach via a tube. This can take some adjusting to and can put a strain on the parent as they try to cope day to day and adapt to the child's needs. The mother of the three year-old describes this:

“The hardest thing is learning to accept it. Learning to accept that she needs a lot of attention, nappy changing and feeds, especially when they tube-feed and still in nappies, helping her dress and undress.”

Young children can also experience social difficulties post-transplant. The mother of the five year-old describes how adapting to such difficulties has required patience and acceptance that the child may not progress, but that over time her son has progressed far beyond her expectations:

“If we got invited to a party or somewhere when he first came out of isolation, he'd be very clingy to me. Not hysterical, but he wanted to stay with me and he wouldn't want to join in, and I thought ‘God, will that ever change?’. ....and now, just slowly but surely that changed. We just kept taking him. I think your fear is ‘well, I'm just not taking him’, so we just kept taking him....what I learnt to do was accept that he might just sit with me and that's fine, and then slowly but surely..... and now it's children he wants, and now it's his friends he wants, and he talks about wanting to go round to his friends, and we've come a massive full circle...It's probably taken two years.”

Regarding learning disabilities and education, the mother of the twelve year-old male describes the difficulty she experiences accepting her child's learning disability when she had higher expectations:

“I was worried about him being mentally handicapped, basically, and [the Consultant] said ‘oh no, we’ve caught it early enough’. But obviously he has got a lot of problems, so I think I kept thinking he’s going to catch up faster than he has. I think that’s a little bit of a disappointment.”

Conversely, the mother of the three year-old female explains how she is prepared to accept that her daughter may not go to mainstream school due to her learning disability:

“I’m hoping to [send her to mainstream school], but if she needs different, I’ll have to accept that. I will accept it, it’s whatever’s going to be best for [my daughter]. I wonder whether the mainstream school reception is going to be too much for her.”

The mother of the five year-old male draws attention to the fact that she previously wished her son to go to mainstream school but how she has subsequently accepted that he is well placed in a special needs school:

“At the moment, my expectations are that he will stay possibly at the special needs school for a while now because it suits him. If by being there and being encouraged and being safe he develops enough to warrant integration then I will be all for it, but it’s not something I outwardly wish for now, which I did two years ago.”



### **5c. Balancing the care of the ill child with the parenting of well siblings**

During the bone marrow transplant parents were required to spend a significant amount of time away from home and had to develop routines and schedules so that the needs of both the hospitalised child and other family members at home could be met. In some cases, families had to temporarily move home to be near the hospital where the child was being treated. This was extremely disruptive to usual family routines and was particularly difficult when there were other children at home. The mother of the seven year-old male explains how during the BMT her other two children felt neglected:

“I think that the other two kids probably think that mum and dad spent more time with [my son] when we were doing hospital and things, going backwards and forwards, than what they got. They don’t resent him and they understand why, but I think they think that [my son] does get more time, more of our time than what they get.”

The mother of the twelve year-old male explains how difficult it was for their other son during their affected son’s BMT, particularly because he was the affected child’s twin and also a young infant at the time:

“[The BMT was] very difficult [for our other son], because he was feeling it because we weren’t with him at times, and he was seeing us coming and going. It was a very unstable period for him. I think for a long time he was very scared of us going, even right up to a few years ago he hated the fact if we weren’t there... So I think it did affect him quite a bit.”

After the BMT, difficulties can continue to arise and parents strive to balance the care of their ill child with that of the child’s well siblings. The mother of the three year-old explains how she makes a concerted effort to treat her two daughters the same despite her affected daughter evidently requiring more attention in terms of care:

“As soon as you get in, after school I always go to the shop or the garage and buy them a book each, you know, a magazine book like

Telly Tubbies or Tweenies, I get them both the same. So I have to make them both the same, but alright [my younger daughter] still needs that extra attention.”

The father of the eleven year-old female explains how their unaffected child has her own ‘extra’ activities that are separate from those geared around their affected daughter, which cater for her own personal interests and abilities, and allow for her own personal development:

“[Our younger daughter] gets plenty [of attention] too. She doesn’t get left out by any means. She gets lots of extra stuff, she’s in karate club and she goes to piano lessons, all these things that [our daughter] doesn’t do. [Our daughter] won’t play the piano because of her fingers, and karate club’s beyond her too, so [our younger daughter] gets her own things.”

Similarly, the mother of the twelve year-old boy expresses concern that her healthy son will feel responsible for his brother. She explains how she feels it important that he has his own interests and friends and develops his own identity that is distinct from that of his brother affected by MPS IH:

“[Our other son] is very good with him in many ways, but I never want him to feel burdened. I kind of want [our other son] to have his friend to himself, because there’s this worry that he’s going to always have to have [his brother] in tow, and that’s where I’d like him to get some friends.”

She goes on to say that despite her healthy son accepting his brother’s illness, she tries to normalise everyday life experiences to help him cope:

“I think he accepts it all. He gets very sad at times, because he wants [his brother] to be like him, but we do try to keep life as normal for [our other son] as possible.”

Similarly, the mother of the sixteen year-old female explains how she tries to make family life as normal as possible for her unaffected children:

“You have to compensate with your children, and you just make it part of their normal life. To them it’s a normal thing that’s going on. Although they know other children aren’t quite like that, you have to try and make it like the normal goings on, which is what I try and do now. Well you can’t, but you try!”

And the mother of the twenty-four year-old male explains how she has tried to treat her son affected by MPS IH as normally as possible, and in fact to overcompensate slightly by giving more attention to her healthy daughter:

“I suppose I really let him be normal. One thing I would say, I tried to put my daughter first to keep her even really, to keep her equal, because there is the tendency if you’ve got a disabled child for your life to revolve round them and you don’t do the best for the other children.”

Again, the mother of the twenty-four year-old female describes how she treated both her children as individuals and met both their needs regardless of whether or not one was sick and the other well:

“[My son] had needs and I fulfilled those, and [my daughter] had needs and I fulfilled those, there was no putting them in different compartments.”

## 6. COGNITIVE APPRAISAL

<b>Sub-Themes</b>	Positive reframing of the child's illness
	Positive reframing: Emphasising the child's abilities and strengths
	Normalisation
	Defining the illness as a meaningful experience: Personal and family development
	Defining the illness as a meaningful experience: Ascribing meaning to the child's presence in the world
	Seeing the child as vulnerable

A number of parents used positive reframing to interpret their child's condition and the situation they are in. For example, most of the parents emphasised their child's achievements beyond their expectations despite the children being significantly disabled and developmentally delayed. Again, despite their disabilities, the parents rejoiced at their child's presence in the world and survival through the BMT. They highlighted their strength of character and hardiness as a result of their experiences, and highlighted how the experience of having a child affected by MPS IH and going through the devastating and traumatic experiences associated with it had enhanced their relationships, enriched their family lives, and led them to develop enhanced personal qualities. They also gave special or religious meaning to their child's presence in the world, claiming that they were 'meant to be', affected by this condition for a reason, and that they have touched lives.

The tendency for families to define the illness as a meaningful event has been demonstrated by previous research (e.g. Folkman, 1997; Wright and Kirby, 1999), as has the recognition of special qualities and strengths in the child and other family members (Hill, 1994; Eiser, 1994), and the feeling that the experience has provided family with the opportunity for growth (Summers et al., 1989). In the situation where a child has survived against all adversity, families have also been found to give special meaning to the child's place in the world and believe their survival to be fated (Mayer, 1982). The ability to define illness as a meaningful event and to put things in a religious context has been related to better family adjustment and better coping (McCubbin and

McCubbin, 1993; Venters, 1981), and the use of positive reframing and the optimistic definition of events as coping strategies (Hill, 1994) have been found to contribute to the maintenance of family unity (Venters, 1981; McCubbin, 1984). Normalisation is another coping strategy that parents of chronically ill children have been found to employ (Eiser, 1990; Monsen, 1999), which was also highlighted in the present study. Parents separated the ‘normal’ aspects of their child’s life from the illness-related aspects when talking about their activities, behaviour, and characteristics. They often used the word ‘normal’ to describe them and defined them and their lives in individualistic terms as opposed to by their MPS condition. Normalisation is suggested to be aimed at minimising stigma and creating a sense of control (Eiser, 1990).

#### **6a. Positive reframing of the child’s illness**

For the most part, the parents interviewed appraised their child’s illness in positive ways. They expressed their gratitude for their survival through the BMT and for their presence in the world, and despite their continued difficulties they greatly valued the opportunity they had to save their child’s life. Despite her son’s significant disabilities, the mother of the five year-old male speaks of the rewards her son has brought her and her husband and how their decision for him to undergo BMT has allowed him to develop into an individual who has exceeded their expectations:

“...The rewards we’ve got from [our son] and the things he’s doing that we never dreamed he would do are fantastic.....I don’t regret it for a second, it was definitely the right decision, I don’t doubt that for a minute. All I think is if he hadn’t have had that transplant he would not be the little boy that we’ve got today. We would never have met the [son] we’ve got now, we’d never know him.”

Similarly, the mother of the seven year-old male describes how thankful she is for the BMT and how it has dramatically changed her son’s future and given him the chance of life. She goes on to describe how seeing a child like her son post-BMT would help parents with the uncertainty they experience at diagnosis and leading up to BMT:

“I have never regretted it, and with every day that I see him, I just, oh well, I cannot believe the changes that that bag of blood can do, it’s amazing.”

The mother of the eleven year-old sees the BMT as being given the chance of a future. The downward comparison she makes with families who do not have the opportunity to have a BMT indicates that she considers herself and her child to be fortunate:

“You’ve got something to work for knowing that when the bone marrow transplant works, then you’ve got a future. That future is uncertain I think, but there is a future there that isn’t the black future that people who don’t get the opportunity have”.

The mother of the twenty-four year-old describes how she lost a part of herself through the trauma of the diagnosis and BMT, but that she doesn’t regret the decision due to the rewards her daughter has brought to her life:

“I think it was only about ten years ago that I realised what an effect it had had on my life, and I think it took a part of my life that obviously I’ll never get back and didn’t at the time realise I was losing. Just a part of you is taken it’s so traumatic. But even so, knowing everything, I know I would still have made the same decision, because of the joy and happiness she’s brought us in her way.”

## **6b. Emphasising child’s abilities and strengths**

When describing their children many of the parents pointed out their strengths and highlighted the progress they had made beyond their expectations. The mother of the five year-old male describes how her son has vastly exceeded her and her husband’s expectations in terms of his communication skills and highlights how he can now function socially outside of the family:

“Two years ago, in terms of his speech and his communication, he had very little means of communication or he could communicate with

immediate family but nobody else. And he can communicate with anybody now! They might have to ask twice and some of his friends don't fully hear him, but in terms of communication he's gone way beyond any expectations we ever had. We didn't think he'd ever speak."

Similarly, the mother of the seven year-old male tells of her expectations being surpassed by her son's abilities:

"He's in mainstream school. He has a full-time support assistant. I would never have imagined this in my wildest dreams, I really wouldn't."

She also highlights her son's strengths and strength of character:

"He's a wee fighter, he never gets stuck with anything at school or at home. If there's something he can't do he'll just struggle on and work it out himself, he's very independent, he wouldn't get anybody to do anything for him, which is good, I think he needs that wee bit of fight there. He's just a wee superstar!"

Similarly, the father of the eleven year-old describes his daughter's resilience and strength of character as a result of her illness:

"She doesn't let her disabilities affect her. She's got a good tolerance of pain, which is probably because of what she's been through. For example, she'll fall over and cut herself and she'll just say 'I've cut myself, whereas her sister will cry for ten minutes.'"

The mother of the twenty-four year-old male describes her son's strength in the face of adversity:

“He’s very confident, very sure of himself, even with all his problems, being in a wheelchair and everything, he’s quite happy to go off on his own when we’re out at the shops.”

She attributes his attitude to his easy going nature and strength of character rather than to his lack of awareness of his disabilities:

“I don’t think he feels as though he’s losing out on anything, or that he’s deprived of anything. It’s not because he’s not bright enough to think that way, I think he’s just an optimistic, easy-going, accepting sort of person. He doesn’t seem to be frustrated by his lack of abilities.”

### **6c. Normalisation**

In describing their children and expressing their views about the condition and their children’s futures, parents often used normalisation. The mother of the seven year-old male describes how she does not see his MPS condition as something that should define him and hold him back from living a normal life:

“.....he’s got Hurlers and it’s a pain in the bum, but he’s not disabled, he’s not any of these things, he’s just [my son], which is the way we’ve brought him up to be. I don’t want him using an illness for excuses or anything like that. It’s just something he’s got and he’s got to live with it.”

The father of the eleven year-old female also describes his daughter as not having special needs and explains how he treats her no differently to how he would treat her if she were not affected by MPS IH:

“I actually don’t consider [my daughter] as a special needs child, I just consider [my daughter] as [my daughter], she’s just my daughter and that’s all there is to it. I don’t even think that she might have a shortened life, I don’t even think of anything like that, I just think of her as my daughter and we just get on with life. I don’t give her any



leeway, she's growing up as a normal child, and that's all there is. The bone marrow transplant was an opportunity to actually do that."

The mother of the twenty-four year-old female similarly describes her daughter as being 'normal' and having needs that needed to be fulfilled just like any other child:

"I always thought of her as normal. No matter what she was, no matter what was wrong with her, she had needs and I fulfilled those needs and never thought of her as different."

The mother of the twenty-four year-old male normalises her son's limitations by describing them as 'shortcomings' and 'different' rather than less than optimal. She also considers that disability should not hold an individual back in life:

"His shortcomings to him are not shortcomings, they're just different maybe to other people, but that shouldn't stop you from doing what you want to do..... He calls himself a stud-muffin. If you say he looks nice, he says 'of course, I'm God's gift!'."

Many of the parents, when describing their children, their lives and the activities that they participated in, separated the 'normal' aspects of their child's life from the illness-related aspects. The mother of the five year-old describes her son's behavioural difficulties but attributes them to his disabilities. She draws attention to his happiness however, and highlights this as the most important outcome they could wish for:

"He has his 'special needs moments' as we call them, but a lot of that is down to his level of understanding, his deafness, and the problems he has anyway with all of that. But generally he's a very happy little boy, and I couldn't have wished for more than that basically."

She goes on to highlight the 'normal' aspects of his life, where he is socially competent, has independence from the family, and lots of friends:

“They’re always in here, he loves sharing his toys, he plays on the street with friends round the corner, I can leave him at other people’s houses with kids and he just goes off and you don’t see him for dust.”

The mother of the seven year-old also describes her son as being happy and having lots of friends, and similarly to the mother of the five year-old male, separates this aspect of the child’s life from the illness-related aspect of being in and out of hospital:

“I think he’s quite happy with his life now to be honest. He’s got a huge circle of friends, he’s not in and out of hospital all the time, yeah I think he’s quite a happy wee guy.”

Similarly, the father of the twelve year-old male highlights his son’s happiness and fun-loving nature:

“He’s fun loving, mischievous, energetic, bossy. He’s a very happy boy. He enjoys himself.”

The father of the eleven year-old female emphasises his daughter’s sociability and participation in different activities:

“She’s very sociable..... She goes to all the after school clubs, brownies, she’s with the drama club.”

The mother of the twenty-four year-old male describes her son as being sociable, confident, and independent:

“He meets friends, he goes off. He’s just been away for the weekend and he’s always happy to go off, he makes friends, it doesn’t matter that he doesn’t know anybody when he gets there.”

She goes on to separate the individual from the condition when talking about her son’s adaptive functioning limitations in terms of communication:

“He keeps his own thoughts very much to himself. Even at school – ‘what did you do at school today?’. ‘Not a lot!’. I don’t know if that’s just a boy anyway. Having one of each – my daughter and I sit and discuss all sorts of things, she’ll tell me what’s been happening with her day, and when she came home from school she told me everything, who said what and who did what, but [my son] has always kept that [to himself].”

When talking about their child’s future some of the parents described the aspirations they had for them, including independence and relationships. The mother of the seven year-old male speaks a lot about her son’s future independence from the family and how important she feels it to be. She also explains how her son’s condition should not prevent him from achieving this and separates his illness from his right to live a full and independent life:

“Independence, I think is really important. They have to be independent, fully. [My son] has been skiing and everything. I just want them to grow up being happy and independent, and not need me, because I think that’s really important, especially for [my son], because if he does make it be an adult, which I hope he does, I want him to be out there with his brother and sister having the time of his life, like he should be. It’s his right to be, Hurlers shouldn’t stop that.”

She has strong views that her son should experience all the things that a healthy teenager should when he reaches that age:

“I can’t wait, to be honest. I think it would be great to see him driving a car, and hopefully going to college or uni or something, and just being a typical teenager, which I think he would be anyway.”

She goes on to describe his aspirations regarding a future career and explains that with a little modification due to his limitations she feels he is likely to fulfil his dreams:

“His dad’s a fireman, and for a long time, ‘I’m going to be a fireman mummy’, but he’s changed it, he wants to be a policeman now, and we keep teasing him. ‘Well you’d better hurry up and grow then if you’re going to be a policeman’. So, yeah he is starting to chat now about what he’d like to be and things like that, the typical boy things, the fireman, the policeman. So yeah, he will. I’m sure he’ll do something like that, maybe not on the fire engine, I can’t see that happening, but doing something.”

The mother of the twenty-four year-old male describes her son’s sexuality as ‘normal’ and attributes his lack of a relationship to him not currently having an interest in anyone:

“I think he probably would [like a relationship], although he’d probably be very reluctant to spend his money on them. He hasn’t got anybody that he’s keen on at the moment. So, I know his feelings are normal. There was a girl that actually liked him, and we were a bit disappointed that he didn’t pursue it, because her parents had a nice big house with their own swimming pool.”

#### **6d. Defining the illness as a meaningful experience – personal and family development**

The majority of the parents interviewed reported that the experience of having their child and going through repeated traumatic episodes had provided them with the opportunity for growth and had lead to the enhancement of family relationships. In particular, parents talked about how their experiences had brought them together as partners. The mother of the three year-old female describes this:

“Between us we seemed to get closer, because we knew we had to be there for [our daughter].”

The mother of the five year-old male describes how due to the serious nature of their child's condition they did not fall out over trivial matters as their friends did, and how their experiences have brought them closer as a couple:

“We do everything together. We share all the care for [our son]. It's strange, I know when you have children it changes a relationship and I remember when we first had [our son], the first year, a lot of our friends would fall out with their partners over a lot of different things, just because everything had changed, and because we had something so major to deal with we actually didn't, and it's probably actually brought us closer together.”

Similarly, the mother of the twelve year-old describes her and her husband's mutual understanding and how their experiences have brought them closer together:

“[My husband] is my best friend. I think it has got us closer, because I know that [my husband] is the only one that understands, and he knows that I'm the only one that understands, and we can only rely on one another....”

She also goes on to say that their experiences have enhanced them as individuals:

“I think in a way, without sounding pompous, it's made us better people, because we're more considerate.”

The mother of the seven year-old male describes how her son's illness has lead her to value her children more which has resulted in a more enhanced family life:

“There's a lot more family time than probably what there ever would have been. Because if you've got healthy kids, you do take it for granted, we did, we did. But we don't now, we cherish everything.”

The mother of the twenty-four year-old male explains how due to her son's illness they have done more as a family and been able to experience things they wouldn't normally have:

“We've probably done more as a family because of [our son] than we would have done.... We've been to Florida about 5 times! Maybe we wouldn't have even been once if I just had two normal kids. ....I'd say we did everything possible as a family. There's nothing we haven't done, because of [my son]. There's probably a lot of things we have done because of [my son], as I say, going to Disney and all that sort of stuff.”

#### **6e. Defining the illness as a meaningful event - ascribing meaning to the child's presence in the world**

Three of the eight families interviewed held strong religious or spiritual beliefs and felt their children had been born and brought into the world for a significant reason. They felt their child's presence in the world was meant to be. They also felt they had been blessed or that someone or something spiritual was looking after them when events fell into place smoothly or when fortune shone on them. The father of the eleven year-old female describes the 'miracle' of finding a bone marrow match for their daughter:

“They found somebody who was almost a match, and then the miracle happened between the first transplant, somebody suddenly appeared on the register who'd been on the register about two weeks who was a ninety-plus percent match.”

He goes on to describe how after the transplant his daughter became seriously ill with pneumonia and that the treatment given was expected to destroy her bone marrow. He draws attention to her survival against all adversity and gives special meaning to this:

“Then she came out and went back into intensive care and they said 'well unfortunately the stuff we had to give her to kill off the pneumonia also kills off the bone marrow', but it didn't, so there you

go, she was meant to be, and she was, she went through all that and a lot more since.”

The mother of the twelve year-old male gives special meaning to her son’s presence in the world and to him being affected by MPS IH:

“I’m very protective towards him, I just want him to fit in, and I know that there’s a place for him, because I believe there’s a place for everybody. He’s here for a reason and he’s got this condition for a reason.”

She goes on to describe how she feels they are being looked after by God or in a spiritual sense:

“I’m quite religious and I believe that we have had a lot of help. I believe that we are getting a lot of help, strange things have happened. [My husband] had a strange experience and kind of said to me ‘it’s going to be alright’, and he was right. That was when he was first diagnosed.”

Similarly, the mother of the twenty-four year-old male gives special meaning to her son’s presence in the world and to his illness:

“I look back and I think the things that all fell into place seem strange, as if he was meant to be here. I don’t know how to describe it without it sounding silly. I’m a practicing Catholic, and I just think that we were meant to have [our son]. The lives that he’s touched since he’s been here.”

## 6f. Seeing the child as vulnerable

As well as using positive reframing to interpret their child's illness and the situation they were in, the parents interviewed also drew attention to their perception of their child as vulnerable. The majority of the parents made reference on more than one occasion to their child's mortality, having limited time with their child and thus making the most of the time they had, and their uncertainty regarding the child's longevity. It was evident for some of the families that the perception that their child might not be alive the following year has impacted upon the way they have psychologically managed the condition and subsequently parented the child. Perceptions of child vulnerability are conceptualised as anxious cognitions about the child's health and susceptibility to illness and death (e.g. Forsyth et al., 1996; Green and Solnit, 1964). Increased perceptions of child vulnerability have been linked to a number of outcomes for children that could interfere with their adjustment (e.g. Allen et al., 2004; Anthony et al., 2003; Maiman et al., 1986; Spurrier et al., 2000). The mother of the five year-old male says:

”.....and it might still be, and *we don't know if he's going to outlive us or not.*” (Emphasis added)

The mother of the seven year-old male explains how she did not expect her son to survive the transplant and that living to five years old he had exceeded her expectations:

“I didn't think [my son] would survive to be five. *I never ever imagined that he would have made that age.....even with the transplant*, when I went to Manchester to do his transplant I didn't think I'd bring him home again, I really didn't.” (Emphasis added)

When talking about her son's future independence she adds the caveat ‘if he makes it to be an adult’:

“I just want them to grow up being happy and independent, and not need me, because I think that's really important, especially for [my



son], *because if he does make it be an adult*, which I hope he does, I want him to be out there with his brother and sister having the time of his life, like he should be.” (Emphasis added)

She also draws attention to her uncertainty regarding her son’s longevity when talking about how she copes:

“[The single most important thing that’s helped me to cope is] just the will to want him to live. Making sure that his life, *no matter how long or short it is*, is a good one. Get up every day to fight and make sure he’s here tomorrow.” (Emphasis added)

The mother of the twenty-four year-old male emphasises the fact that healthy as well as sick children can die, but she similarly draws attention to the fact that there is a great deal of uncertainty regarding longevity with this patient group and it therefore has an impact on their life:

“[My son] has lost friends at school, they’ve died for different reasons, and of course that makes me think about what’ll happen when [my son] goes *and I don’t know when that will be. I think there’s always that hanging over us*, but then friends have lost children getting run over or whatever. *It’s just because we know that the possibility is there.*” (Emphasis added)

The mother of the twenty-four year-old describes how from her daughter’s diagnosis there has always been a question mark over her survival and that this uncertainty prevails:

“From the day that she was diagnosed I suppose, every day was a bonus, she had Hurlers, she was therefore not expected to survive, she had her first transplant that failed, she had a second transplant, there were no promises, *I mean they still don’t know how long she’s going to live*. So we learnt in the very beginning...to live one day at a time.” (Emphasis added)

She goes on to say:

“Yes, I was always aware and I suppose I still am of making the most of the time you’ve got, *because you don’t know when that might finish.*” (Emphasis added)

## 7. STRESS AND COPING

Sub-Themes	Perceptions of stress
	Taking one step at a time
	The use of different coping styles and strategies
	Coping self-efficacy

With regard to perceptions of stress, the parents in the present study spoke of both psychosocial and illness-related stressors. In terms of psychosocial stressors, parents spoke of the experience of seeing their child in pain and distress, and seeing them frightened and disturbed by experiences. These were related to the BMT, routine hospital visits, and major surgery. One mother also described the distress experienced witnessing her daughter’s social difficulties at school. With regard to illness-related stressors parents made reference to characteristics of their child in terms of their fussiness and need for routine, and to the commitment to care. The mother of the sixteen year-old female spoke at length about her daughter’s constant presence in the home, her dependence on the family, and lack of motivation to do anything outside of the house, and highlighted this as a source of stress. The mothers of the two young adults both described how their commitment to care over many years was taking its toll and how they had no freedom or social life. Functional care strain in terms of helping their child with activities of daily living was not highlighted by the parents as a source of stress. This may be because they do not perceive it as being stressful due to the positive perceptions they have of their child’s situation and their ability to cope, thus moderating the effects.

While both illness-related and psychosocial stressors have been related to parent adjustment by previous research (e.g. Ievers and Drotar, 1996; Thompson and Gustafson, 1996), other factors have been highlighted as moderating the effects of such stressors. Intrapersonal factors, such as self-efficacy, self-esteem, and feelings of control, social-ecological factors, such as social support and family functioning, and stress processing factors such as appraisals of situations and coping processes have been found to moderate the effects of stress on personal adjustment (e.g. Wallander and Varni, 1998; Wallander et al., 1989). Individual appraisals of situations, such as the degree to which mothers' perceive the child's illness to impact on the family has been highlighted as a mediating process through which illness or functional severity may lead to parental psychological problems (e.g. Wallander et al., 1989; Ireys and Silver, 1996; Lustig et al., 1996). Mothers' appraisal of disability-related stress has also been strongly related to her maladjustment (Noojin and Wallander, 1997) and parent illness-related stress has been shown to be exacerbated by lack of knowledge about a condition (e.g. Ievers and Drotar, 1996; Brewer et al., 2008).

### **7a. Perceptions of stress**

As mentioned above, each of the families interviewed remarked on different experiences that they found stressful. For some it was the on-going day-to-day stress of caring for a child with disabilities or a particularly stressful period of adjustment that their child went through. The mother of the eleven year-old female makes reference here to her daughter's need for attention:

“[I would say] her constant demand for attention [has been the most difficult aspect of living with her condition].”

The mother of the seven year-old describes how difficult it is coping with her son's need for routine:

“[He's very fussy about what he eats], very. Right down to the bowl he uses and the plate and the knife and fork and the cup, but he'll only eat [certain things], and trying to make that a bit different every night is awful.”

The mother of the sixteen year-old female describes how difficult it is to cope with her daughter's dependence on the family, her lack of motivation, confidence, and social competence:

“I think emotionally it is very, very hard to deal with, and obviously they can't do anything socially so they're always in the house, and that is really, really hard. ....she needs to have a life apart from the family, because this is all she does every single day of her life. She needs to have something else in her life to talk about, to be positive about because otherwise she would be here doing what she does all the time, watching television. And she needs that encouragement, that push to do things.”

Both mothers of the young adults describe how their ongoing commitment to care has become increasingly difficult to cope with. The mother of the twenty-four year-old male explains:

“I suppose the biggest difficulty is our freedom, mine and my husband's. Everything is geared around [our son] and at twenty-four your lives shouldn't be geared around your children. So that's upper most in my mind.”

The mother of the twenty-four year-old female explains how the uncertainty regarding her daughter's longevity has had an impact on her commitment to care, and that had she known what to expect, she may have parented her differently:

“[My husband] and I have never had a social life since [our daughter] was born, and I suppose now it's getting more difficult to cope with all those things that I'm beginning to realise perhaps what I missed out on. I think that that's probably it, I think maybe I've been over-committed. Probably if I'd been told when she had her transplant she's going to live 40 years and that will be it. Maybe I would have done things differently, but you just go day by day.”

She points out that:

“[The most difficult aspect of living with my daughter’s condition] is the commitment I think, and especially as I get older.”

Other stressful events highlighted by parents include those surrounding the transplant, hospitalisations, and surgery. The father of the eleven year-old female describes his experience of his daughter’s illness following her bone marrow transplant:

“I sat for days just watching the monitors and willing them not to stop, because that’s the only thing I had, she wasn’t breathing, she was on a ventilator.”

He goes on to describes the effect the stress of the BMT had on his wife:

“My wife was pregnant at the time [of the BMT] as well and she lost that baby through stress. That baby was unaffected, but she lost it at 6 months through stress. That baby was lost between the two transplants.”

The mother of the twenty-four year-old female describes the transplant itself:

“It was very hard, it was really very hard, and I suppose seeing [my daughter] going through what she did.....I remember when they started taking her immune system away she had to have about eight different drugs, and I remember a nurse literally pinning her on the bed and my daughter screaming her head off and these drugs being literally poured down her. You know, it was awful, I just wanted to say ‘get off my baby, leave my baby alone!’”

The mother of the seven year-old male describes her experience of seeing her son go through serious orthopaedic surgery:

“He would bite me, he would kick me, he would just get so distressed...and [my son] is not an aggressive child and he’s never hit anyone in his life, but at that time he’d just get so distressed he won’t come out of his wheelchair, he’s absolutely miserable, he won’t eat, it’s just awful, it’s horrible to see him so sad and so frightened as well.”

The mother of the sixteen year-old female describes her experience of seeing her daughter endure social difficulties at school:

“When she started secondary school it was a different ball game, because although the children were told that she’s disabled and needs a bit of help, because of [my child] not being social she couldn’t start a conversation still with anybody and she would wait for them to talk to her, and even then she can’t really carry on a conversation, or it’s inappropriate conversation. [So] because of that the children just didn’t want to interact with her. So that was very hard.”

### **7b. Taking one step at a time**

Examples of coping processes were highlighted by parents, in particular parents’ tendency to take things one step at a time. A few parents spoke of their preference to live one day at a time rather than think too much about the future. This, one father indicated, prevented them from having expectations about their daughter’s progress that could be dashed. Instead, this approach enabled them to appreciate their child’s small achievements in a piecemeal fashion. Previous research has illustrated how parents of chronically ill children use the expression ‘living one day at a time’ (Ray, 2002), illustrating the non-linear and ongoing process of adaptation. It is seen as a practical response, which allows families to deal with problems as they arise and focus on managing that situation (Hill, 1994; Jerrett, 1994; Atkin and Ahmad, 2000), and offers protection against ‘engulfment’ (Twigg and Atkin, 1994) and overwhelming thoughts about an uncertain future and a lifetime of burden (Atkin and Ahmad, 2000). The father of the eleven year-old describes how this approach helps him to cope with his uncertainties regarding his daughter’s future development:

“I think our day to day approach is the right one. I think if you have too many expectations and too many ambitions you might get disappointed. So we work day by day and [our daughter] achieves something each day and it’s great.”

The father of the twelve year-old explains how they do not put too much emphasis on the future indicating that he feels it is too full of uncertainties:

“It’s sort of a one day at a time thing with us. We’re planning for our futures and things like that but living as it is, we just live for today, plan for tomorrow.”

The mother of the twenty-four year-old more explicitly describes how due to not knowing how long her daughter might live learning to take one day at a time has helped her to cope:

“The minister of our church when [my daughter] had her first transplant really taught me to live one day at a time, and that was the best lesson I ever had, and I think.....You were always expecting that maybe next year she wouldn’t be here, so you just live for today and didn’t look to the future.”

### **7c. The use of different coping styles and strategies**

Parents described a variety of strategies they used to help them cope during stressful times, including taking time out, talking to friends and partners, and distraction. Some went on to describe how they felt they coped in different ways to their partners, which they explained caused difficulties. Previous research has suggested that individuals use a variety of coping strategies depending on the situation and the response that is needed, and that one coping style is not necessarily more effective than another. However, within partnerships coping styles that are incongruent with one another can have implications for the kind of support couples can offer each other, for effective communication and parents’ ability to cope. One of the fathers interviewed for this

study described how his wife liked to talk about her problems while he preferred to go away and sort things out privately, and one of the mothers described how her husband blocked things out, found it difficult to accept his daughter's illness, and generally did not cope well. This, she explained, resulted in her taking the emotional as well as physical burden of caring for their daughter for over two decades.

Previous research has illustrated how parents' differential ways of communicating such as these can create difficulties, particularly for mothers (Pelchat et al., 2003), how such communication difficulties can effect parents' ability to cope with their child's illness (Whyte, 1992), and how differential coping styles can impact on the kind of support parents can give to and receive from one another (Bristol et al., 1988). Since the family's vulnerability to stress can be reduced by the establishment of a support system within the family (Holaday, 1984) and family functioning increased by the quality of the marital relationship and parents' ability to share the burden (Venters, 1981; McCubbin, 1984), it is vital that differential perceptions of parenting stress and coping are better understood amongst mothers and fathers of children and young people affected by MPS IH post-BMT, not least because the quality of the family environment has implications for child psychosocial outcomes (e.g. Finney and Bonner, 1992; McCubbin et al., 1993). The mother of the five year-old explains how, during the transplant, it was difficult to talk to her husband about a particular issue for fear of upsetting him and how she found it helpful to talk to a counsellor on this occasion:

“There was a time when we were going through the worst of it, I think it was just somebody to blame. I had to be angry at somebody, so there was a bit of time when I felt angry at [my husband's] family. I didn't want to talk to [my husband] about it because we were going through enough without me slagging his family off to him. So I had a few weeks of counselling, which I found really useful, just someone impartial that I could let off steam to.”

She goes on to say that she also talks to friends as a means of coping day-to-day:



“Day in day out now I have a good cry every now and again, but my biggest way of coping is to talk about it, which is why I talk endlessly to my friends.”

The mother of the three year-old female also finds the support of friends important:

“You know that thing where family or friends will phone up and say “what are you doing, are you coming out with us?” Or “do you want to go into town?”, so I’m always doing something, and I think that’s what keeps me going.”

Distraction and keeping oneself busy is another form of coping that parents use. The mother of the seven year-old describes this:

“You just fire into something else and get busy again and don’t have time to think.”

Similarly, the mother of the twelve year-old male explains how focussing on other people’s problems helps her to cope with her own:

“I think another way of me coping is that I trained to be an aromatherapist, so now I’m helping people with their problems and that’s how I cope, because I can forget about my own problems and help them.”

During particularly stressful periods, like when her son is undergoing surgery, the mother of the twenty-four year-old explains how she has to distract herself in order to cope:

“[When he’s having operations] I have to go off and take my mind off things completely, go shopping, walk round like a zombie, but I can’t just sit and wait, I have to have a mission, I always have to be doing something to keep my mind off it.”

Although the source of the stress is often related to the care of the child and having to see them experience pain and difficulties, a means of coping with that stress can also be focussing on the rewards the child brings and on ensuring their wellness and survival. The mother of the five year-old male describes how her child's character helps her to cope:

“The hardest thing is [my son], but the most amazing thing is [my son]. He'll always end up doing something every day that just has you in absolute stitches.”

Similarly, the mother of the twenty-four year-old male describes how she couldn't cope without her son's positive personal characteristics:

“I couldn't cope with it without [my son] being the way he is, his personality gets us through everything.”

The mother of the seven year-old male also describes how her will to do the best she can for her son enables her to cope:

“[The single most important thing that has helped me to cope is] just the will to want him to live. Making sure that his life, no matter how long or short it is, is a good one. Get up every day to fight and made sure he's here tomorrow.”

Similarly, the mother of the twenty-four year-old female describes how the will to want her child to survive drove her to cope through the transplant:

“It's experiences I wouldn't wish on anyone, horrendous to go through, but you do at the time, because you're almost on a some sort of wave in a way. You're just fighting for this goal, for your child's life, and you just do it.”

Other parents use withdrawal as a means of coping. Both parents of the twelve year-old boy describe how they withdrew from the world in order to cope with their son's diagnosis and how this has made it easier for them to cope. The mother explains:

“When this all happened I think we just went into our own little world a little bit, and I’m not sure that we’ve ever completely come out of it.”

The father explains how this has enabled them to cope:

“I think sometimes it’s just the easiest way through it, just to shut the doors and get on with it.”

Two of the mothers describe how taking time-out helps them to cope day-to-day. The mother of the three year-old explains:

“I don’t actually get low, low. I have my moments when I like to put the telly on and put a video on [for the children], and go in the kitchen and have a cup of coffee and a cigarette.”

The mother of the sixteen year-old says:

“You’ve got to have that bit of space just to keep you sane. Chill out and have a cup of coffee and sit and watch what I’m going to watch, you have to have that bit of space.”

Furthermore, within families individuals can use different coping strategies and styles, which may either complement one another or be incongruent. The father of the eleven year-old female describes how his daughter's diagnosis led family members to use different strategies in order to cope with the information:

“I spent about three hours going through a dictionary trying to work out what all these words meant, my wife spent the day trying to find help. I think it was her that found the MPS Society. I think [my daughter's] grandmother cried, her granddad went out and dug the rose bed for

about five hours, people just did their own thing just to sort their own mind out. It was horrendous.”

He goes on to explain how he and his wife cope in different ways, suggesting that the support they give one another may be out of tune with their needs:

“I’m not a big communicator. I’m a believer in going away quietly sorting out your problems, and once you’ve got them sorted out, come back and deal with them. Whereas I think my wife likes to talk about problems.”

#### **7d. Coping Self-Efficacy**

Some of the parents also highlighted how they felt at times that they did not cope well with their child’s illness and its related stressors or how their coping self-efficacy hit peaks and troughs just as did the adaptation process. Intrapersonal factors such as perceived self-efficacy have been highlighted as important in buffering the negative effects of stress (Bandura, 1982). For many parents of chronically ill children, feeling that they can manage the condition on a day-to-day basis has been reported as being beneficial to them (Atkin and Ahmad, 2000), as has feeling confident about their problem-solving abilities, which has been linked to the use of more active coping strategies and more positive feelings toward parenting (Noojin and Wallander, 1997). The organisation of care and other such normalising tactics have also been found to help family adaptation (Krulik, 1980), to reduce the impact the illness has on the child (Holaday, 1984), and perceived self-efficacy has been related to parents’ reports of adaptive family functioning (Kazak et al., 2004). Understanding how manageable parents find their situation when they have a chronically ill child, and how well they feel they cope, is important if appropriate support is to be given. The mother of the three year-old female describes the commitment to care that her daughter requires, but states how she feels she copes well:

“I find that because she’s not like another normal child, she needs that attention – she can’t get in the bath on her own, she can’t dress herself, she can’t climb into a car on her own, she needs a lot of support, *and I*

*think I do it quite well....the feeding and nappy changing. The only time that I don't really need to do anything is when she's in bed, unless she's having a bad night."* (Emphasis added)

The mother of the five year-old however questions her coping self-efficacy at times, but normalises this tendency by stating that nobody feels they are coping well all of the time:

"I don't know how well I cope or I don't cope. Generally we cope very well I think, and we do, we get through it and we do a lot. We function as a family and we go out and do loads, so yeah we probably do cope, but then sometimes you feel like you're not, but then I suppose everybody does to a certain extent."

She goes on to describe how she can be quite hard on herself when she finds it difficult to cope, but manages to justify it by recognising that she has more to cope with than most parents:

"I sometimes go 'God how come everyone else can do this and that, how come everyone else can have four, five or six children around on their own and I can't?' I thought I'd be this great supermum, and I beat myself up about it, and it's like 'well no, I could have five or six kids as long as [my son] wasn't there."

The mother of the seven year-old describes how she does not feel she copes well but puts on a façade to protect her son:

"Inside I probably [don't cope] very well, outside I try to get on with it as much as I can and hope that I'm alright to support [my son] through it. I'm always concerned that if he sees me frightened or worried, that it's going to rub off on him. I really panic, but I really try my hardest not to let him see that."

The father of the eleven year-old female describes how he feels he copes well by that his wife has difficulties which has resulted in serious consequences:

“I’m fine, my wife is alcohol dependent now. She has to go to a clinic and get sorted, but she’s in a mess. It’s the way she coped, but it’s not the way to do it is it? Because now it’s out of control.”

The mother of the twenty-four year-old female explains how she has found it very difficult at times to cope despite her faith:

“I’ve always had quite a strong faith, and I think the one bit of teaching that everybody says to me that I’ve had probably over the last ten years is, ‘The Lord never gives you more than you can cope with’, and I think ‘well he really tries me sometimes’, because you go down to the pits and it’s very difficult to crawl back out.”

## 8. SOCIAL SUPPORT

Sub-Themes	Valued sources of support
	Sharing the experience with others who have ‘been there’
	Unhelpful support
	Lack of support

The social-ecological resistance factor of social support was highlighted by all the parents as vital to their ability to cope. This reflects previous views that being able to draw on social support is an important coping strategy (Lazarus and Folkman, 1984). Valued sources of support described varied from partners, who shared the care of the child, offered appropriate support, and understood how the other spouse felt, to friends who offered understanding, a break, someone to talk to or just company, and extended family members such as grandparents who offered invaluable support, understanding and respite. A great deal of research has highlighted the importance of support to parents of chronically ill children, and how it serves to buffer the effects of stress. With regard to the marital relationship, spousal support is cited as being the most important

type of support for mothers (Ray, 2002). It is reported as being predictive of maternal adjustment to childhood chronic illness (McCubbin and Patterson, 1983) and seen to protect mothers against stress, especially when fathers highly value child-rearing activities (Nagy and Ungerer, 1990). It is also highlighted as an important factor in family adaptation for both mothers and fathers (Goldberg et al., 1986; Grant and Whittell, 2000). The support of grandparents has also been found to be valuable for both mothers and fathers (Oka and Ueda, 1998; Pelchat et al., 2003), and social support in general has been found to be beneficial to parents of chronically ill children and be related to their health and well being (Schwarzer and Leppin, 1989; Warfield et al., 1999).

### **8a. Valued sources of supports**

Parents were asked about their sources of support and all described valued support in their lives from a variety of sources, including spouses, friends, and members of their extended family. The mother of the five year-old male describes her husband's support as being in tune with her needs and their partnership as supportive and complementary:

“[My husband] is a completely one hundred percent hands-on dad, he just takes him out, he'll take both of them out, we share everything. If one of us is feeling a bit tired or is not feeling particularly patient, the other one will be. It seems to just somehow work, if one of you is not coping the other one is, and it always just seems to work out that way.”

She goes on to explain how her husband's support and the cohesiveness of their family have enabled her to cope with her son's illness:

“The single most important thing that has helped me to cope is my husband, being very much together as a couple really, and a family, and having an agreed view on what we want for [our son].”

Similarly, the mother of the twelve year-old male describes her husband's support as invaluable and describes their relationship as complementary and mutually supportive:

“[My husband] in particular is quite a strong person, he’s very positive, and I think at times when I’ve fallen over he’s picked me up and maybe the odd time that he has then I’ve done the same.”

The mother of the sixteen year-old also explains how the support of her husband is important, particularly when she is having a difficult time with her daughter:

“So I sort of say to him ‘oh she’s been really playing up today, can you hear her, she’s having a strop’ and he’ll come in and he’ll have words with her, so he will back me up on stuff like that. So yeah it’s just the backing up thing, I think that’s quite important because....he knows what she’s like and he knows that it gets to a point where I’m going to grit my teeth.....because I’ve had it from when she comes in from school and he comes in from work and gets the tail end of it.”

The mother of the twenty-four year-old also describes her husband as a supportive parent:

“He’s always been hands-on as a dad. When he came home from work he would take over. He couldn’t understand fathers that didn’t, because how do you bond with your child and get to know them?”

Parents also spoke of friendships and the important role friends play in supporting them. The mother of the three year-old explains how she was worried about how her friends would react to the news of her daughter’s diagnosis, but having shown them information leaflets about the condition she has found that they continue to offer support when it is needed:

“I’ve got a lot of close family and a lot of close friends. When we first found out about [my daughter], I wondered how my friends would accept it, and so just showed them the booklets. Instead of explaining it to them or trying to, I just got the booklets and said there you are, and just passed them round. They’re really, really helpful. If there was



anything I needed, or I needed to go out for an hour with just me and my older daughter or anything, they will offer to watch [my daughter].”

The mother of the five year-old male explains how her friends are an important source of support, how they show understanding towards her son, have a relationship with him, and help her to make decisions regarding his care:

“If [I have a problem with my son], I’ll say to my friends ‘what do you think we should do, what should I do?’ And they will support whatever I decide and that is a massive way of how I cope. Because they understand him so well because they’ve spent so much time with him, I can talk to them, they can give me their advice, or I know that they’ll support whatever I decide.”

Similarly, the mother of the sixteen year-old has friends who have a long-standing relationship with her daughter and thus understand the difficulties that her condition can bring. This she explains helps her to talk about any issues she has:

“....a couple of my friends, they’ve known [my daughter] since she was a dot, so they know what she’s like and they know that she can be grumpy and stuff, so yeah I do have a moan to them and have a chat, but any questions and things I will ring the MPS.”

Again, the mother of the twenty-four year-old describes how she and her husband have a large circle of long-term friends who understand their experiences and with whom they feel safe:

“[We get our support from] friends, lots of friends. We have friends that we’ve had for years and years. We feel secure in the people around us, and in ourselves, my husband and I. No problems for ourselves as a couple. Friends that we go away with and spend time with.”

Parents also talked about the support of extended family members and how this has helped them to cope. The mother of the five year-old male describes how the support of

her parents has been invaluable and how the knowledge of their unfaltering support enabled her and her husband to make the decision to have another child:

“We’ve both got brilliant families who are very supportive and have just supported everything we’ve done, and been there. My mum and dad, [my husband’s] parents still work, but my mum and dad are constantly there, and that’s why we went on to have [our daughter] because we knew we would have the extra support, because that’s needed.”

The mother of the eleven year-old also speaks of her family and how their support provides her with some respite:

“Having family back-up has been important. I have enough [family around me] to have some relief from the stress. My mum will take the girls for a weekend occasionally.”

#### **8b. Sharing the experience with others who have ‘been there’**

Being able to meet other families of children affected by MPS IH who had undergone BMT and who had been through similar experiences was also highlighted as a valued source of support. These experiences were described by parents as comforting as they felt understood and did not feel the need to explain themselves. They also helped parents to understand the possible outcomes of BMT prior to their child undergoing treatment, helping to alleviate some of the uncertainty experienced at this time. The benefits of such experiences have been previously reported. For example, being able to talk to other parents in similar situations and who have ‘been there’ has been reported as being beneficial for coping (McCubbin, 1984). It allows parents the opportunity to gather and share information and increases understanding of the illness (Holaday, 1984). Parents can also feel part of a community, find sources of emotional support, develop greater mastery and control over their situation, see more meaning in their situation, and feel more accepting of their child’s illness or disability. This can have positive consequences for child psychosocial outcomes (Soloman et al., 2001).

One of the tertiary centres that specialises in MPS diseases in the UK holds a joint clinic for bone marrow recipients affected by MPS IH which is held approximately twice a year. Parents attend with their children and get the opportunity to meet other families. The MPS Society also organises conferences and social events, and gives families the opportunity to attend other related conferences in the UK and abroad. Again, this is an opportunity for families to meet and socialise with one another. The mother of the five year-old describes how attending the joint clinics enabled her to have clearer expectations of the BMT and to feel more confident about the outcomes:

“I think joint clinics are a really good idea, because we met a couple of other children who were probably what [my son’s] age is now with their school uniforms on and chatting and doing things. Ok they might have had a back brace, they might have had the hearing aids, but they were lovely, and chatting, and real personalities, and you could see that for their families [the bone marrow transplant] had worked and it was well worth it. So I think, for me, that was really important.”

The mother of the seven year-old explains how helpful she would have found it to meet a child who had undergone BMT when she was making the decision for her son to undergo treatment:

“Oh, if I had seen a child like [my son] when I went through transplant it would have been amazing. It wouldn’t have changed how I felt, it would probably have given me a bit more confidence to do it, because I really was petrified.”

The mother of the eleven year-old also explains how seeing both treated and untreated children affected by MPS IH helped them to make the decision for their daughter to undergo BMT:

“We met a little girl from Blackpool who had been through a bone marrow, and although she was a bit small and bit shy, we were quite happy for [our daughter] to be like her. So we did see the not treated

and we did see the treated, and we decided that [our daughter] should be given every chance.”

The father of the twelve year-old male describes how comforting it felt being around people who had been through the same experiences:

“When we went to that conference it was really nice, because people understood exactly what you were talking about, and in fact, you almost didn’t have to say anything, because they understood, they’ve been there, done it, and got the tee-shirt.”

### **8c. Unhelpful support**

Research has also highlighted however, that support offered can be of the wrong kind or out of tune with the recipients needs, and can have negative consequences for parents. For example, mothers and fathers are often differentially impacted upon by childhood chronic illness and can respond in different ways (Gray, 2003). This can have implications for family functioning and intra-familial supportive behaviours, and in turn for child psychosocial outcomes. As is illustrated by the mother of the twenty-four year-old female in the present study, mothers can often take responsibility for the day to day care of the child, and if one partner copes more effectively than another or family support is lacking or of the wrong kind it can have negative consequences in terms of stress. The lack of family support such as this has been reported as a strong predictor of stress amongst mothers of chronically ill children (Margalit et al., 1989; Atkin and Ahmad, 2000). Equally, fathers can experience adjustment difficulties but may have different concerns to mothers. This again can have implications for the kind of support that is needed (e.g. Margalit et al., 1989; Lillie, 1993; Pelchat et al., 2003). This again draws attention to the differential experiences of mothers and fathers in terms of stress and coping, and reinforces the importance of learning more about them, particularly amongst parents of children and young people affected by MPS IH post-BMT, to gain further understanding of how they may impact on both parent and child adjustment.

Whilst most parents in the present study had one or more sources of support which helped them to cope, some felt that the support offered was not adequate or in tune with their needs, while others even had experiences where support was explicitly withdrawn or not offered. This was from immediate as well as extended family members. As mentioned above, the mother of the twenty-four year-old female describes how, despite her husband appearing to be supportive, she has taken the burden of caring for their daughter onto her shoulders:

“[With my husband] it’s always this ‘oh it’ll be ok’. Everything to [my husband], whatever happens, whatever disaster there is, ‘it’ll be alright, don’t worry about it, it’ll be alright’, but somebody in that scenario has got to do the basics, has got to do the work to make it alright, and I think that someone has always been me.”

She goes on to say that her taking on the responsibility for the day-to-day care of her daughter has been the result of an unspoken contract between her and her husband:

“I said to my husband ‘I can’t remember that you ever said to me, this is going to be hard for you will you do it?’ It was just ‘you will do it’, not that he said ‘you will do it’ [like an order], but it was just expected of me [to care for our daughter].”

She adds:

“I suppose [my husband has] always been there for me, but he just can’t cope with it.”

The mother of the twelve year-old male describes how her brother might ask about her daughter but that he is doing nothing more than paying lip service to her situation and failing to properly support her:

“I can’t say that I’ve had a lot of practical help. My eldest brother has always been in the States, but he will ring up and always ask how [my daughter] is. And maybe I would start telling him about what was

happening medically, and you could almost hear him go to sleep on the phone, and I think ‘well why bother asking’.”

#### **8d. Lack of support**

As well as unhelpful support from spouses and other family members, parents also spoke of a complete lack of support. One set, the parents of the twelve year old male, described how their requests for support had been flatly rejected by family members, how they showed no understanding of their situation, and how they had never been offered a break by family members. Previous research has highlighted how families can cut themselves off from family support because they feel that others do not understand their situation or feelings or as a means of protecting their child (e.g. Speice et al., 2002; Todd, 2007), which can have implications for adjustment (Mechanic, 1977). This was evident with this particular family. In the following extract the father of the twelve year-old male explains how his request for support and understanding from his father was rejected:

“I wrote to my father trying to explain what it is that we go through all the time, having to go to [a city] and that, and he wrote me back and told us to stop wallowing in self pity, don’t write again. So that’s why we say the boys have got no grandparents, although my dad’s still alive, he’s not a part of their lives at all, and he lives with my sister in Scotland, so they’re not a part of it either.”

His wife goes on to explain how as a result of this they no longer expect support from others and have learnt to rely only on themselves:

“I felt that we hadn’t had the support from people that I wish we had have done. [My husband’s] family, for example, no support whatsoever. [My husband’s] favourite saying is, ‘don’t expect anything and then you’re not disappointed’ and I think that’s how we live really...”

She adds to this by describing how she and her husband have never had offers of support until very recently:

“It wasn’t until earlier this year that someone actually offered to have the boys for a weekend. We’ve never had that, it was the first time. Most people have got grandparents and you can get rid of the kids for a weekend, but we’ve never had that.”

## **9. LACK OF UNDERSTANDING**

The feeling that others’ lacked understanding of their experiences was also highlighted by a number of parents. This included never being offered a break by family members, which was interpreted as failing to understand the experience of having a disabled child, and feeling that others do not understand when trying to explain one’s feelings. Parents also talked about their upset at immediate as well as extended family members’ lack of understanding about their child’s behaviour and limitations. Such experiences have been previously reported as causing distress to parents of chronically ill or disabled children (e.g. Pelchat et al., 2003; Ray, 2002) and as leading families to isolate themselves from family support as previously mentioned (e.g. Speice et al., 2002; Todd, 2007). The role that social support plays in alleviating the stress associated with having a chronically ill child is a complex one and not fully understood. However, if the support given is deemed to be effective, it has been found to be of benefit to parents (Rappaport, 1984). The effect that perceived support and understanding can have on parents’ adjustment to having a child affected by MPS IH post-BMT is therefore important to explore in more depth. The mother of the five year-old male describes how members of her husband’s family felt uncomfortable with her son’s disabilities and how she felt angry at their lack of understanding:

“I think a lot of [my husband’s family] found it quite hard to start with, but now [my son] is a bit more of a character, now he’s talking, now he can communicate with them, they’re trying a lot harder to communicate back, but I think they were a bit scared. I don’t think [my husband] was, but I think I was a bit angry at first that they didn’t make as much of an effort as I’d have liked, but I realise now that

everybody deals with things differently, and it was their way of dealing with it.”

The mother of the twelve year-old explains how people’s lack of understanding of their situation has led them to stop trying to seek support and to explain how they feel:

“I’ve never been the sort of person to ring up somebody and tell them our problems, and yet we get a lot of people ringing us. I did try once to tell my sister-in-law how I felt, because she often phones me up with her problems, but she just didn’t understand, and I didn’t bother any more.”

She goes on to explain her reasons for not giving MPS information out to friends at the time of their son’s diagnosis and describes how she felt people were not particularly forthcoming with offers of support and ultimately could not understand how they felt:

“I often thought that maybe we would have [given MPS information to our friends], but I think the sort of friends that we had at that time would not have really understood. Don’t get me wrong, people were kind and if we’d have said ‘can you help?’ they probably would have done, but we didn’t find people were very ‘now look, if there’s anything we can do....’ I think it was beyond a lot of people, they just didn’t understand.”

Similarly, the mother of the twenty-four year-old female describes how she has never been offered support and how this is the result of people not understanding what it is like to have a disabled child:

“[My family] never say to me ‘ooh, you should have a break, we’ll come and stay with [your daughter]’. So a lot of people don’t understand what a commitment it is, what it actually involves, ‘oh yes she’s got a disabled child’ or whatever but they don’t realise what that entails.



The mother of the sixteen year-old female explains how there is a lack of understanding of her daughter's behaviour within the family. She describes how her older daughter has experienced jealousy towards her sister in the past but despite this passing continues to fail to understand her sister's condition and the behaviours related to it:

“Although the jealousy has gone, [my older daughter] doesn't really understand things with her. She's quite good with talking to her now, she'll try and talk her into things and she'll try and explain things with her, but she hasn't got that understanding of why she does things differently sometimes.”

The father of the eleven year-old describes a lack of understanding of his daughter's needs within the school environment. He feels that his daughter has required educational support rather than just a care assistant:

“She's got a very, very short attention span and she's very easily distracted and she needed somebody with a bit more formal training in education than somebody who could wipe her nose and take her to the toilet.”

Similarly, the mother of the five year-old male describes her son's original placing in a mainstream nursery as being unsuitable as they did not understand his needs and how to appropriately deal with his limitations:

“Oh school are brilliant, and they have such a wonderful balance. They completely understand his needs and what he can and can't do as an individual, but they completely understand what they expect and what he is capable of. And that's what I found actually, it got to a point at the mainstream nursery where he got away with 'oh it's just him being him', so he actually did get away with murder. The difference with [the special needs] school is, yes he's got these needs and they have to be taken into consideration, but he also needs to [sit down and be taught]. So it's a better balance I would say.”

## 10. WANTING TO PROTECT THE CHILD

The issue of protection was talked about by many of the parents interviewed, which covered a number of areas. One mother described how difficult she found it to exercise normal parenting behaviours such as disciplining her son as she found it difficult to forget the difficult time he had had and the challenges that faced him. Others talked of wanting to protect their child from the pain and fear associated with aspects of the illness itself, from danger, the outside world, and developing autonomous behaviours to the point of over-protection. These issues have been previously written about and attention has been drawn to mothers' tendency to become ensconced in their parenting role when they have a chronically ill or disabled child, wanting to protect them from harm and take their pain (Pelchat et al., 2003; Twigg and Atkin, 1994). Over-protection has also been reported in mothers of chronically ill children, which can be detrimental to the child's psychosocial development and the development of autonomous and self-help behaviours necessary for adult life (Anderson and Coyne, 1991; Holmbeck et al., 2002). Due to the uncertainty that parents of children affected by MPS IH post-BMT feel, this is an issue that requires further exploration, particularly in relation to child psychosocial outcomes. It is also an issue that needs to be brought to the attention of health professionals so that appropriate support can be given. The mother of the three year-old describes how she is generally more protective towards her daughter affected by MPS IH:

“I do [think I've changed as a parent] yeah. I'm not as laid back as I was.... I'm more protective. With [my daughter], the slightest cough I think 'should I take her to the doctors'. I'm more protective.”

The mother of the five year-old male explains how difficult it is to instil normal disciplinary measures when she has the knowledge of the difficulties her son has been through and will continue to have to endure:

“You've always got in the back of your head everything else that he's been through and everything else he's got to go through, and you try and be neutral, but when he's doing something and you've got to tell

him off, and he's having a paddy or whatever, you can't help but think 'well in six months time he might be having his knees stapled'."

She goes on to say:

"It's very hard to tell [my son] off, well we found it quite hard initially because he's been through so much. It's not his fault, he's done this, this, this, and this, but for his sake in the long term he's got to have these boundaries, but we've found it hard."

The mother of the seven year-old male describes her desire to protect her child from the consequences of the illness itself after her son went through a particularly traumatic surgical experience:

"[After his spinal surgery] I just wanted to close all the doors and keep him safe, and really do think that he didn't feel safe anywhere, and felt he doesn't even trust his mummy, because his mummy hurt him too. I think he had a really hard time and it took him a long time to get over it."

The mother of the twelve year-old male describes how she wants to protect her son from the prejudices of the outside world:

"Anybody that is slightly different, some people will single them out, but yeah, that is my concern. I just want to protect him and maybe I'm a bit overprotective at the moment, but I just want to try and protect him from anybody trying to hurt him."

The mother of the eleven year-old female explains how her daughter has been allowed to do very little unsupervised, and continues to go to few activities without being accompanied. In this instance the mother's reference to playing outside refers to the immediate vicinity of the house on the parents' property, and indicates her concerns for her child when she is unsupervised and away from the home:

“She does, she’ll go and play outside. I think really it’s only Brownies that she actually goes to at the moment that I leave her there. With regard to going to school trips, yes, now, but in the past I’ve gone along with her as well.”

The mother of the twenty-four year-old describes how as a result of her perception of her child’s vulnerability she has perhaps over-protected her daughter throughout her life. She describes how she has tried to take her pain and her worries and feel them for her instead:

“We didn’t have tantrums or anything like that. I don’t know, maybe I just nurtured her through everything. I do feel that I took a lot of her pain, her worries, I was always there for her, and maybe, in hindsight, too much, because I think she’d be a more independent person now if I hadn’t done that. But it comes back to this, ‘well we might not have her next year’.”

She goes on to describe her concern about her daughter potentially embarking upon a sexual relationship and indicates her perception that her daughter is vulnerable and that she is in need of protection in such a situation:

“Have I put her off having any relationship, have I stopped her having any relationship, because I say ‘all men are useless’? And I’ve been quite glad that she has that feeling in a way, because I really don’t know how I would react if she had a relationship.”

The mother of the twenty-four year-old male explains how she continues to feel protective towards her son and worries about him when he goes out, though she does not prevent him from acting independently:

“Oh, it’s still hard [for me when he goes out]. I still hate it, I still want to follow him, but I have to pretend I don’t care. No, it is difficult. But as I say, the mobile [phone] makes a big difference really, so he can keep in touch.”

## 11. PARENTS PERCEPTIONS OF CHILD PSYCHOSOCIAL WELL-BEING AND DEVELOPMENT

<b>Sub-Themes</b>	Psychosocial functioning: The need for routine
	Cognitive and adaptive functioning
	Social competence
	Behavioural difficulties
	Children's understanding of their illness/disability
	Children's adaptation to illness-related stressors

A number of child characteristics, adjustment, and developmental issues were explored by parents. Most described their child as needing fixed routines and how they would become distressed if these routines changed in any way. One mother described her daughter as exhibiting obsessive behaviours. Most parents also described their child as having limited communication skills and at some point in their lives experiencing social difficulties and social inhibition and withdrawal. For the most part this followed the BMT and major surgical procedures. Following this however there was some variability in parents' descriptions of their child's social competency, personality, and independence. Some were described as continuing to be inhibited and withdrawn, with one, the sixteen year-old female, become more so as she got older. Others were described as being very sociable, enjoying the company of friends, and going out to socialise away from the home without parental supervision. No major externalising behavioural difficulties were reported, other than those related to medical procedures and hospitalisations. However, these were reported as becoming less severe as the children got older and adapted to medical procedures becoming a normal part of life. With regard to self-perception and the children's awareness of their condition, again parents reported different feelings. Some parents described how their child was aware of their difference to others and their limitations and showed displeasure at this. The mother of the twelve year-old male described how her son demonstrated his awareness of his limitations by his behaviour and becoming withdrawn following failure. Other parents described their child as not considering themselves to be disabled, not seeing

their difference as a short-coming, or of having no concept that the aids they use such as hearing aids, back braces, and wheelchairs are out of the ordinary.

When asked to describe their child, most of the parents gave a balanced account of their personalities, describing both positive and negative aspects of their character. For example, the mother of the three year-old female points out that she can be both loving and have tantrums:

”[My daughter’s] personality is loving. She has quite a lot of tantrums. Otherwise, in herself, she’s just a loving little girl, without her tantrums.”

Similarly, the mother of the five year-old male describes her son as being both happy and carefree but also stubborn:

“He has two very different sides to him... he’s generally very happy, very carefree, he loves to do things, but then on the absolute opposite extreme, if he doesn’t want to do something he can be very stubborn.”

In terms of psychosocial adjustment, parents drew attention to difficulties their children experienced with their adaptive functioning in terms of communication and social competence. They also highlighted their children’s need for routine. However, for the most part they described their children as reasonably well adjusted. The exception to this was the mother of the sixteen year-old female who drew particular attention to her daughter’s negative demeanour, comparing it to when she was younger and a much happier individual:

“She’s quite grumpy a lot of the time. .... Whereas when she was smaller she was quite happy a lot of the time, but now it seems to be... she doesn’t show her emotions, let’s put it that way, she doesn’t show happiness.”

### **11a. Psychosocial functioning: The need for routine**

In terms of routine, all of the parents highlighted their children's need for routine and their likelihood to become significantly distressed if unexpected changes were made. This included diet as well as activities. This perhaps indicates an adjustment difficulty and suggests that the children and young people may feel insecure when anything other than the norm is presented to them. This is illustrated in the following extracts. The mother of the five year-old male describes her son's reaction to change and novel situations:

“Anything that is not ongoing and routine is very difficult..... He is completely freaked out by something he doesn't know.”

The mother of the seven year-old male makes the same point:

“His routine never changes. ...That routine is automatic and if I decide one day 'oh we'll go down the shops' or whatever, 'we'll have tea out' or something, it throws him. He can't cope with that, he just doesn't like it.”

The mother of the sixteen year-old female describes how routines have become more fixed the older her daughter gets:

“Come fifteen and sixteen, she's very set in her ways, she doesn't like it if anything changes.”

She goes on to describe how this kind of behaviour becomes obsessive at times:

“Sometimes she's quite obsessive about certain things, ie (sic) maybe closing the cat-flap at a certain time, making sure the cat is where it should be at a certain time, having a certain food on a certain day. She's obsessive about doors being closed, she doesn't want the animals out. She [verbally] repeats things quite a lot.”

Similarly, the mother of the twenty-four year-old female explains how her daughter gets very upset by any change:

“...she likes everything to [be in] order..... [and] she still very much likes her lifestyle as it is and doesn't like any change. ...any change at all completely upsets [her]. The slightest little thing and she just takes a long time to absorb it, why it's happened, why things have to change. She's very happy, as long as there aren't any of those changes.”

### **11b. Cognitive and adaptive functioning**

All of the children were developmentally delayed in terms of speech, albeit to varying degrees. Most of them were therefore described by their parents as having limited communication skills, particularly in relation to the articulation of feelings. The mother of the three year-old describes how her daughter communicates her leg pain to her:

“If she's hurting, she can tell me that her back's hurting or her knees are hurting, she'll start rubbing her knees....”

The mother of the seven year-old describes her son's communications skills as limited but improving:

“[He can articulate his emotions] slightly. You get 'I'm sore' or 'I was sad today', 'I fell out with my friend'. I don't think he's a hundred percent but it's starting, he is starting to.”

The mother of the eleven year-old female explains how her daughter's ability to communicate happy feelings is limited:

“She won't so much say what's made her happy, but if something has happened to make her sad then she will come and tell us that, yes.”



Again, the mother of the sixteen year-old female describes how difficult it is to gauge her daughter's emotions:

“No [she can't articulate the way she feels]. You have to guess. Obviously you know when she's really, really, really upset because she'll get so frustrated that she'll cry and then you know she's upset about whatever it is, she's been told off or you say to her that something wasn't nice”.

Similarly, the mother of the twenty-four year-old female describes her daughter's limited range of emotional expression:

“....everything to [my daughter] is on the same plateau – if she has to have an operation, if she has to go to Disneyland, everything is on the same level, which is weird, but that's how she accepts things. And she doesn't really get excited....it's just satisfaction, 'yes, this is what I want, this is what I like' and that's it. So I suppose on the other side as well, she doesn't show [sadness]. She likes watching old films and I say 'oh you're not watching this sloppy rubbish again are you!' and she says 'oh it's sad', but that's about as far as the emotion goes.”

Parents made reference to their children's cognitive impairments and communication difficulties and the impact this had on their ability to interact with others socially. The mother of the five year-old male explains how her son's limitations have led him to interact with younger children:

“He played at the mainstream nursery but he began to play with the younger ones slightly, and that's kind of happened with some of my friends as well. He plays with his peers but when they get onto more complicated games he plays with the younger brothers and sisters, and they all tend to muck in and play together but he veered towards the younger ones.”

The father of the twelve year-old describes his daughter's tendency to play with younger children, indicating that by doing so she protects herself from feelings of inadequacy:

“She doesn't have many friends of her own age, she tends to relate to much younger children. If we go on holiday, for example, she won't play with the 10- and 11-year-olds, she'll play with the 6 or 7 year-olds. I suppose she feels like she's king of the castle when she's with them, whereas it's the opposite way round when she's with her peers.”

### **11c. Social competence**

Many of the parents described their children's social difficulties, particularly in young childhood, thus not long after the bone marrow transplant. The mother of the three year-old describes her daughter's timidity in social situations and her unwillingness to interact with other children:

“She won't really mix until she gets to know them and then it could be..... Like she went to a party, she was only at school two days and this little boy invited her to his party. It was from two 'til four and she didn't move from my knee, she just would not mix with the other children. They were dancing and she'd do all the actions on my knee, but she wouldn't leave my knee.”

The mother of the seven year-old male describes her son's similar reaction when he was in young childhood, though she goes on in her interview to describe her son's social development since this time:

“He was absolutely petrified of everyone. His only friends up until he was probably about 5 was [his older brother] and [older sister], that was it. He would play in the room with lots of children round about it, but he didn't want to be with them, he didn't want to join in with their games or let them join in with him. He liked the noise and having

people around about him that he knew, but he wouldn't play with them. So I think it has affected him quite a bit, then it had."

Parents' descriptions of their children's social competence varied amongst those with older children. Some of them were described as being socially inhibited some as being unable to interact socially, and others as being able to enjoy active social lives. The father of the twelve year-old male describes him as lacking in confidence in social situations:

"He gets shy with strangers, he gets a bit nervous and he hasn't got a lot of confidence when it comes to [talking to people he doesn't know]."

His mother goes on to explain how she is unsure of his understanding of friendship:

"Whether he initiates any [social interaction] I'm not too sure, because I think he doesn't really understand having a special friend. I think he is popular because I think the other children want to play with him, but whether he understands about having that special friend [I don't know]."

The mother of the sixteen year-old female describes her daughter's difficulties interacting with her peers:

"She's confident with adults, but adults that she knows of or knows. She's not particularly fond of starting conversations, she finds it really, really hard to start conversations, even with an adult. She won't even attempt it with her peers, unless she really knows them."

She goes on to explain the limited nature of the friendship she has at school with one of her peers:

“She’s only got one friend, but the little girl who pushes her around there and stays with her at playtime and stuff, she’s able bodied but she’s a little bit slow with talking.”

The mother of the twenty-four year-old female describes her daughter as introverted and indicates that she has always been so:

“She’s quite introvert, and I don’t mean by that she sat in the corner and wouldn’t mix in with the other children, she did, but she didn’t really get involved in a lot of school activities.”

Similarly to the sixteen year-old female, she adds that her daughter has had an acquaintance throughout her school and college careers but that their relationship is extremely limited:

“Her friend who she had been with almost all through her school life, and they keep meeting up at various colleges and now he’s at the day centre, but apart from saying hello to each other, I don’t think they natter too much, I wouldn’t say they were bosom pals.”

In contrast, the mother of the twenty-four year-old male describes his as socially competent and able to make friends with peers who are not disabled and to choose his friends:

“He’s got able-bodied friends that he goes out with, that he keeps in touch with. They go to the cinema or down the pub or whatever. He used to spend a lot of time with [my daughter] and her friends, but through his choice he stopped hanging around with them.”

### **11d. Behavioural difficulties**

While withdrawal, inhibition, and other internalising behaviours were described by parents, externalising behavioural problems were not raised as major issues by parents, with one exception. As previously discussed, the sixteen year-old female was described by her mother as having obsessions and compulsions, using inappropriate language, and being physically aggressive. Other behavioural problems were described as the result of frustration, a lack of understanding and confusion or as being directly related to stressful events or experiences, such as major surgery, or pain. The mother of the three year-old describes her daughter's tantrums:

“She has quite a lot of tantrums. If she can't get her own way she's throwing herself down onto the floor and stamping her feet, and she's starting biting now, and nipping.”

The mother of the five year-old describes her son's tendency to lash out when he experiences frustration or a lack of understanding in a situation:

“When he's frustrated or tired or doesn't understand something he can hit out occasionally at me and [my husband], but I've never seen him do it with any of his friends.”

The mother of the seven year-old describes her son's behavioural difficulties in relation to hospitals:

“He does have [behavioural problems] with hospitals and things with getting distressed, and he would get distressed in nursery, and at school he went through a long time of checking every cupboard and every room, and I really think he was looking for a treatment room, I really do.”

The father of the eleven year-old describes minor aggressions displayed by his daughter:

“If she doesn’t get her own way, particularly with her sister, she’ll nip her or slap her”

### **11e. Children’s understanding of their illness/disability**

The extent of physical disability experienced by the children and young people in the sample varied. The two youngest children, who were three and five years of age, and the seven and sixteen year-olds were mobile, but used buggies or wheelchairs half of the time or when on long journeys. The eleven year-old walked with no problem and did not use a wheelchair at all, although she became tired or experienced pain by the end of the day. This was also true of the twelve year-old, however his recent knee surgery had left him a little less stable on his legs than usual. The two twenty-four year-olds were both wheelchair-bound and could no longer bear weight. Their learning difficulties also varied, however an objective measure of intellectual functioning was not carried out for this phase of the study. When the parents were asked how their children saw themselves, most described their children as being aware of their disabilities, but felt that they did not necessarily experience difficulties associated with their self-concept as a result. The mother of the five year-old male explains how her son accepts the additional items that come with his condition as a normal part of life:

“He’s always fully accepted his hearing aids, he’s worn them since he was three months, and if you don’t put them in, he asks to have them in. I had to leave his corset off today because he was very chesty, but he’s like ‘mummy, corset’. So he knows that they’re all his things and that he has them, but I don’t think he has any inkling that other children don’t.”

The mother of the seven year-old male describes how her son does not perceive himself as being disabled:

“A couple of weeks ago we were Christmas shopping and I went to go in to the disabled toilet. He’s in his wheelchair, he pulls on the brakes and he’s like ‘what are you doing?’, and I’m like, ‘I’m going to the toilet with you, shy?’, ‘I’m not going in there, I’m not disabled!’”

With regard to learning disability, two of the children were reported by their parents as having an awareness which affected them emotionally. The father of the eleven year-old describes how his daughter becomes upset when it becomes apparent that her younger sister is more academically able than her:

“[She’s got a] younger [sister], who’s very clever, and that doesn’t help sometimes either, because her sister can run rings around her academically.”

He goes on to describe his daughter’s concern that her younger sister might catch up with or overtake her in school:

“She didn’t want to be in the same class as her sister. She always wanted to be in a higher class than her sister. She’d say ‘when [my younger sister] moves up, will I move up as well?’ She asked me that one year, I said ‘of course you will’. She didn’t want her sister to be in the same class as her because she’s the big sister.”

And the upset that is caused if her younger sister demonstrates her superior abilities:

“Put it this way, if [my daughter] is doing her homework with me and her little sister just goes past and shouts an answer out it really, really upsets [my daughter], because it’s *her* homework and her little sister shouldn’t be doing it. Her sister’s very capable and that sometimes is a problem.” [Emphasis original]

The mother of the twelve year-old male describes her son as becoming withdrawn when he is unable to do something:

“[When he can’t do something he gets a bit] withdrawn, although it doesn’t last for long, because we tend to move on and that’s it. It doesn’t last long.”

However, she goes on to say that he will not try again following a failure, indicating a learned helplessness response:

“I think that’s his way of dealing with things he can’t do, [and] it’s pretty much the same at school. I get the feeling from what his teacher said, that if he knows he can’t do [something], he won’t try. It’s just finished, he’s not going to do it.”

The awareness of being ‘different’ from other children was also an issue for some. The father of the eleven year-old describes how his daughter is aware of her difference to others and of some aspects of her disabilities:

“Oh she knows she’s different now. She’s at the age where she knows that..... and she does say sometimes, ‘why have I got my back like this?’ She’s not happy with her back.”

The father of the twelve year-old male explains how his son is becoming aware that his twin brother is more able than him:

“He doesn’t comment [that his brother is better able to do certain things], but I think he is aware sometimes.”

The mother of the sixteen year-old describes how her daughter comments on her difference to others and on other people’s tendency to stare at her:

“She knows that she’s different and that’s the only thing you can tell her if people stare, ‘oh you look a bit different that’s all’..... She knows she’s not normal, she often says that, she does often remark on it”.

In contrast, the mother of the twenty-four year-old female describes her daughter as not having any concept of her disabilities or condition and that as long as her limited needs are met she is contented:



“She’s never kicked her wheelchair, ‘why have I got to be in this? I want to walk’. She’s never said ‘why do I look like this, why don’t I look pretty?’ Never ever, she’s very contented, weird!”

#### **11f. Child’s adaptation to illness-related stressors**

Frequent hospitalisations play a prominent role in the lives of children affected by MPS IH following bone marrow transplant. They go for regular health checks, including blood and skeletal monitoring. Most of the children in this sample have had major orthopaedic surgery at some point in their lives. Adapting to hospitalisation, for these children, therefore forms part of their adjustment to disability. However, hospital visits and surgery can be very distressing for a young child, and can be disruptive to their lives. Parents talked about the distress that their children had experienced with hospital visits, and how over the years the majority were adapting to the visits as a necessary and normal part of their lives. The mother of the three year-old female describes her daughter’s distress on these occasions:

“She cries. She gets herself really upset, but she always has done, so I don’t know if she’s just going to carry on like that. She gets herself really distressed, especially the blood, she had all her vaccinations, it took three of us to pin her down, which is shocking.”

The mother of the seven year-old explains how as her son has got older he has found it easier to cope with hospital visits, and how her ability to communicate with him has helped that:

“If he needs bloods or anything done it would take four or five of us just to get that done, which is horrific, it’s awful. As he’s got older, the last time I was down in Manchester, about two months ago, he was fabulous. He’s getting older now and I can say to him ‘nobody’s going to hurt you this time, you’re going to get pictures done’ – that’s x-rays – or ‘nobody’s going to jab you and you’re not staying, we’re going home this afternoon’ and he can cope with that now.”

The mother of the eleven year-old explains how hospital visits are becoming more difficult for her daughter as she gets older:

“She’s not too keen [on hospitals]. The older she gets the worse it is.  
She doesn’t like needles at all”

The mother of the twelve year-old male, like many of the others, describes how hospital visits in the past were horrendously upsetting, but how as her son has got used to them he has adjusted and accepted them as a normal part of life:

“When he was little it used to take five of us to hold him down for an x-ray, now he’ll lie there quite happily. He’s getting better, he’s getting use to it, I think he just accepts it as part of life now.”

The mother of the twenty-four year-old male explains how her son used to feel anxious about hospital visits, but how that has got easier as he has got older:

“He went through a very bad patch when he was very nervous of hospitals, anyone in a white coat, but as he got older it got easier.”

She goes on to describe how he now relishes in the experience:

“In fact, it now gets to the stage where if they’re putting the needle in, his heads getting in the way ‘cos he’s so busy looking at what they’re doing. And it’s a joke now, ‘do you want me to come with you?’, ‘No, I don’t, thank you!’. ‘I can take it like a man’ he says, ‘us Palace supporters can take it’. He just makes a joke of it.”

These reports suggest that there are differences among the children and young people affected by MPS IH post-BMT whose parents participated in this study, particularly in terms of confidence, social competence, and independence. There are also differences between them in terms of cognitive, adaptive, and physical functioning. This reflects the view that chronically ill and disabled children can experience psychological morbidity and adjustment difficulties (e.g. Koopmans and Lamers, 2000; Lavigne and

Faier-Routman, 1992; Wallander and Varni, 1998, 2000) and problems in peer relationships (e.g. Noll et al., 1991). While there are a number of factors that put chronically ill children at risk of adjustment difficulties however, there are a number of resistance factors that serve as moderating factors and protect them from the negative effects of stress (Wallander et al., 1989), and a great deal of research supports the view that the psychosocial functioning of chronically ill and disabled children is more related to individual, family, and parenting factors rather than to disease or disability-related factors alone (e.g. Lavigne and Faier-Routman, 1992; Wallander and Varni, 1998; Wallander et al., 1989). So that a better understanding of the psychosocial outcomes of BMT for this patient group, and the processes involved, can be reached, it is therefore of vital importance that such outcomes are explored in more depth and in relation to the individual, familial, and parenting factors. It is important also that reports are taken from the children and young people directly where possible, as well as via their parents, so that reliable measures of functioning can be obtained.

### **3.4. Overview and conclusions**

The purpose of this first exploratory stage of the study was to illuminate the issues that were pertinent to parents of children affected by MPS IH post-BMT and to explore their experiences. This was achieved by examining a number of different issues pertinent to parents of chronically ill children as highlighted by the literature reviewed and to those affected by this condition as highlighted by personal communication with health and support professionals that work with this patient group. The results have illustrated many of the experiences of families of children affected by MPS IH post-BMT. This helps to increase understanding of living with this condition and of some of the issues that the affected children and young people may face. A number of super ordinate themes were highlighted: shock and disbelief at diagnosis, no decision to be made about whether or not to undergo BMT, uncertainty, having low expectations of the child, adaptation as an ongoing process, cognitive appraisal, stress and coping, social support, lack of understanding, wanting to protect the child, and parents' perceptions of the child's psychosocial well-being and development. This research can claim originality on the basis of its specific examination of this condition and the

experiences of the families affected by it. Indeed, this is the first study of this nature that has examined the psychosocial sequelae of this condition.

While this qualitative phase of the study provided information in an area that has not previously been researched, and hopefully makes a significant contribution in its own right, no generalisations can be made about other parents of children affected by MPS IH post-BMT and no relationships between individual, family, and parenting factors and child psychosocial outcomes can be assumed, which is the overall aim of the study. Since only parents were interviewed for this phase, which was the study's main limitation, few conclusions can be made about child psychosocial outcomes, whether they experience any marked difficulties, and how, if at all, other factors might impact on that functioning. The experiences of the parents interviewed may also not reflect those of other family members and of the children themselves which may differ. Furthermore, all of the parents interviewed were biological parents of the affected children, which, since the condition is genetically transmitted, could have implications for the issues highlighted by parents and for parent coping. However, this may not be the case for all parents of children affected by this condition who could be adoptive, step, foster, or grandparents. Thus, a further caveat against the generalisation of findings is needed for this reason. A second phase, which employs quantitative methods and involves both parents where possible and the affected children, is therefore necessary in order to explore the experiences highlighted, widen the scope to explore child experiences and development, and to find relationships between variables.

In conclusion then, the qualitative approach used for this phase of the study has highlighted in rich detail the experiences of these parents, which enables a better understanding of the challenges they face and the way in which they view their situation and their child's illness. The sample was selected from within specified age groups so that the experiences of parents of children at different ages could be explored. This reflects the age range of the total population of this patient group in the UK and therefore provides important information about parents' experiences at different developmental stages. This is particularly useful when exploring a progressive condition that is potentially life-limiting and presents families with a great deal of uncertainty. While no generalisations can be made about the wider patient group and families affected by other conditions, the findings here may assist health professionals

and support organisations in their support of families affected by MPS IH post-BMT at particularly difficult times and throughout the child's life. They may also be of relevance to research into the impact other childhood chronic conditions have on the family, and they of course facilitate the development of a robust research tool as they have allowed for the honing of variables.

### 3.5. How phase one of the study informs the second phase

In relation to this study's substantive findings it is apparent that parents of children affected by MPS IH post BMT are impacted upon by both illness-related and psychosocial stressors. Factors that potentially buffer or exacerbate the negative effects of those stressors are also apparent, and include a number of internal and external resources and sources of support, and parental appraisals and perceptions of stress and support and child vulnerability. In line with the disability stress-coping model (Wallander et al., 1989; Wallander and Varni, 1992) a number of risk and resistance factors have therefore been highlighted by this study, which warrant further in-depth exploration with this patient group and their families. The aim of this is to investigate whether or not resistance factors moderate the effects of stress on parent and child adjustment when they are affected by MPS IH post-BMT.

The objectives of the second phase of this study were three-fold:

1. To examine the degree to which intrapersonal, social-ecological, and stress processing resistance factors moderate the effects of psychosocial and illness-related risk factors on parent adjustment.
2. To examine the degree to which intrapersonal, social-ecological, and stress processing factors moderate the effects of psychosocial and illness-related risk factors on patient participant psychosocial adjustment.
3. To assess whether spouses differentially experience parenting stress, coping, and perceptions of child care needs, and whether potential discrepancies in scores or a shared view has implications for parent well-being, and in turn, child psychosocial development by serving as a moderating factor.

In terms of risk factors, both psychosocial and illness-related stressors were highlighted by this first phase of the study, which supports the exploration of parents' experiences of stress in a number of areas, including the child's physical functioning and need for assistance with activities of daily living, child health, child adaptive and cognitive functioning, and child psychosocial outcomes. In terms of resistance factors, which are purported to buffer the effects of such illness-related and psychosocial stressors, the way that parents appraise their situation and the child's illness and cope warrants further investigation, including the beliefs they hold and their perceptions of child vulnerability. These would constitute stress processing resistance factors. Social-ecological resistance factors that warrant further investigation include family functioning, social support, and shared or discrepant views of parenting stress and coping between mothers and fathers. Intrapersonal factors that warrant further investigation include mother's mental health and psychological well-being, and perceptions of manageability. Outcome variables that warrant investigation for parents include parenting stress, coping abilities, psychological well-being, and stress processing in terms of perceptions of child vulnerability.

In terms of child psychosocial outcomes the risk factors that warrant further investigation include health and disability-related stressors, and adaptive and cognitive functioning. Resistance factors include parent stress-processing factors which shape child attitudes and coping mechanisms (e.g. Burlew et al., 2000), including parent's perceptions of parenting stress and their coping abilities, parent's beliefs and attitudes, parents perceptions of child vulnerability, and parent intrapersonal factors, such as maternal mental health and perceptions of manageability. Social-ecological factors that warrant further investigation in relation to child psychosocial outcomes include family environment, parents' shared or discrepant views of parenting stress and coping, and mother's social support. Intrapersonal factors include self-esteem and self-concept, social competence, and variables associated with personal adjustment. The psychosocial outcomes under investigation should include social competency and behavioural outcomes relating to inhibition and withdrawal. Personal adjustment in terms of self esteem, interpersonal relationships and adjustment difficulties also warrant investigation, as does self-image. Since the exploration of child psychosocial outcomes and disease and disability parameters were limited by this first phase of the study, as discussed above, the design of the second phase in terms of the exploration of child

psychosocial outcomes has been partly informed by the issues highlighted by the parents interviewed but also by the literature reviewed in the previous chapter.

It is hypothesised that mothers will experience significantly more parenting stress than fathers, but that they will cope significantly better than fathers with the stresses of caring for a child affected by MPS IH post-BMT. In terms of patient adjustment, it is hypothesised that patient participants will have significantly less well developed adaptive and social skills than a normative sample and that they will exhibit behavioural problems of an internalising nature. It is also hypothesised that psychosocial resistance factors, including intrapersonal, stress processing, and social-ecological factors, will significantly contribute to the variance in both parent and patient adjustment and that they will moderate the negative effects of illness-related stressors on adjustment outcomes.

## **CHAPTER FOUR**

### **QUANTITATIVE PHASE**

#### **4.0. Methodology**

This second phase of the study employed a cross-sectional survey design using a battery of validated questionnaires, demographic and biographical interview questions, questionnaires designed for the purpose of this study, and patient participant cognitive testing. Data were collected in four phases. Firstly through postal questionnaires which were sent to mothers and fathers jointly (where applicable) prior to the researcher's visit; Secondly, via a survey, which was administered by the research to mothers only. Thirdly, questionnaires were administered to patient participants aged seven years and over and all patient participants administered tests of cognitive function. In the fourth and final phase fathers were administered questionnaires over the telephone.

#### **4.1. Participants**

Eligible participants were parents of children affected by MPS IH post-BMT living in the UK. Their children affected by the condition also participated in the study. Forty-four families participated in the study. At the close of data collection in May 2005, the total population of children and young people affected by MPS I post-BMT in the UK was 49. This sample therefore represented 90% of the total population of MPS IH patients post-BMT in the UK. In all 44 cases mothers and one child participated, and in 36 cases fathers also participated. With the exception of one family, families had only one child affected by MPS IH, and this was the child that participated. In the case of the family that had more than one affected child, it was intended to include both children but the mother volunteered only one. The majority of families were recruited via the MPS Society in the UK as described in the section entitled Recruitment in point 2.4 of Chapter Two.



The ages of the patient participants who participated ranged from 16 months to 25 years, the distribution of which is shown in Table 4-1 below. The criteria for inclusion required the bone marrow transplant to have engrafted successfully, and for infants to be out of isolation following their bone marrow transplant. By the close of data collection, two further infants were undergoing transplantation, but were not included in the research.

**Table 4-1 Age of Patient Participants**

Age Range	Male	Female	TOTAL
5yrs and Under	11	6	17 (38.6%)
6-11 years	10	5	15 (34.1%)
12-17 years	2	2	4 (9.1%)
18 years +	4	4	8 (18.2%)
<b>TOTAL</b>	<b>27 (61.4%)</b>	<b>17 (38.6%)</b>	<b>44 (100%)</b>

Forty-four biological mothers of MPS IH post-BMT children participated in the research. Thirty-eight were either married or cohabiting with a partner, 36 of whom were the biological fathers of the couple's MPS child. Six of the mothers were single, having either separated or divorced from the father of their child affected by MPS IH. Thirty-three biological and two non-biological fathers who lived as part of the family also participated. As did one biological father who did not live as part of the family. Three biological fathers who lived as part of the family and were married to mothers who took part in the research chose not to participate. Thus a total of 36 fathers took part in the research. Biological fathers who no longer lived as part of the family were given the opportunity to participate in the research at the discretion of the mothers. Only one biological father in this situation participated. Of the eight biological fathers who no longer lived as part of the family, three "never" saw their child affected by MPS IH, two saw them "occasionally", and three saw them "frequently". It was noted however, that one of the fathers who "never" saw their child was no longer alive, but had not had contact with the child for many years prior to his death. Details of marital status, biological parenthood, and paternal participation are shown below in Table 4-2.

**Table 4-2 Parent Participant Marital Status, Biological Parenthood, and Paternal Participation in Research**

Marital Status	Father Took Part		Is Partner Biological Father		TOTAL
	YES	NO	YES	NO	
Married	28	3	30	1	0
Co-Habiting	7	0	6	1	0
Separated	1	3	0	0	4
Divorced	0	2	0	0	2
TOTAL	36	8	36	2	6
					44

Thirty-nine of the families that participated in the research had other children. Two of the 44 individuals affected by MPS IH did not live at the parental home. One lived in residential care, and the other was an adult who lived independently. Seven families had other children living independently away from home. Details of family composition and birth order of the child/young person affected by MPS IH are shown below in Tables 4-3 and 4-4.

**Table 4-3 Number of Children Living at Home Including Child Affected by MPS IH post-BMT**

Number of Families	Number of Children (inc MPS) living at home
7	1
23	2
9	3
3	4
1	7
1	10

**Table 4-4 Birth Order of MPS Child**

Number of Families	Birth Order of MPS Child
1	Only child
1	Twin/Only Children
11	First Child
19	Second Child
8	Third Child
1	Fourth Child
1	Fifth Child
1	Sixth Child
1	Seventh Child

#### **4.1.1. Sample Attrition**

Two of the 48 families did not take part in the research, as they had been lost to follow-up by the Society. One family had two children affected by MPS IH post-BMT and volunteered only one of these children as previously mentioned; and two families volunteered to take part but were unable to due to their children being significantly unwell at the time of participation. Thus 44 families participated in the research.

#### **4.2. Data collection**

Both parents (where applicable) from each of the 44 families, and their children affected by MPS I post-BMT, were invited to volunteer to take part in the study by letter (See Appendix H). Parents of children aged under-18 years were invited to volunteer on their child's behalf. However, MPS patients aged seven years and over were provided with their own Information Sheet (See Appendix I), and were encouraged to discuss the research with their parents before deciding whether or not to participate. Young adult patients aged 18 years and over were personally invited to volunteer to take part in the research by letter. Again, these individuals were provided with their own Information Sheet (See Appendix I) and were asked to give their own consent. Where adult patients were not able to give their own consent due to learning disability, parents were asked to give consent on their behalf (See Appendix J for the consent form).

Parents were invited to volunteer to participate by returning a tear-off reply-slip in a pre-paid envelope provided. They were provided with an Information Sheet (See Appendix I), which explained the aims and procedure of the study. They were informed that both parents and their child affected by MPS I would be involved in the study if they chose to take part. Mothers from each family were invited to take part in a survey, which they were informed would take no longer than two hours and would take place in their home at their convenience. Fathers from each family (where applicable) were invited to complete some short questionnaires by telephone. MPS patients aged seven years and over were invited to take part in a brief survey, and patients of all ages were asked to participate in a test of cognitive function.

The findings of the qualitative phase of the study and the literature reviewed demonstrated that mothers usually take responsibility for the day-to-day care of the child when they are affected by chronic illness. For these reasons, and for consistency purposes, mothers were chosen as the main participants. However, since the literature and the findings of the qualitative phase also demonstrated how mothers and fathers can be differentially impacted upon by childhood chronic illness and can have differential perceptions of stress and use different coping mechanisms, the inclusion of fathers where possible was also considered important. Due to time constraints however, it was decided to focus the exploration of fathers' experiences on their perceptions of child functional care needs, parenting stress, and coping abilities. The same methods of data collection were used for mothers and fathers. The scores on these measures could then be compared with those of the mothers and any discrepancies between them analysed in relation to both parent and child adjustment. The rationale for administering questionnaires to fathers via telephone was to maximise fathers' participation in the study since, for the most part, fathers were not present in the home during the day (when most mothers completed the survey). Where fathers were in the home at the time mothers completed the survey, they were administered the questionnaires face-to-face. Fathers were routinely administered the questionnaires separately from the mother however, to minimise parents influencing one another's responses.

Once the completed reply-slip had been received, the parents were contacted by telephone and an appointment made for the researcher to visit. The MPS I Health

Assessment Questionnaire and the Patient Medical History Questionnaire were sent to the parents by post, along with an appointment confirmation letter, which they were asked to complete prior to the researcher's visit. The reason for this was to give parents time to gather historical data and to minimise time spent completing the face-to-face survey. Potential participants were given the opportunity to ask any questions they wished about the research. Furthermore, contact numbers were provided in the participant invitation letter and on the Information Sheets, as was an offer to parent and patient participants to discuss the research with the researcher at any time.

Mothers completed the survey in their own home at a time of their convenience. Patient participants were also administered questionnaires and tests of cognitive function at home. These took place either on the same day or one day previous or subsequent to each other. Fathers were then administered three short questionnaires. This was either done in person if the father was present when the researcher visited the family home or by telephone within seven days of the mother completing the survey and child testing. All patient participants were tested and administered questionnaires in the morning. This was done to avoid patient fatigue (following school/college/work) from confounding the results.

Patient participants seven years of age and over were administered self-concept and personality measures verbally by the researcher, and all patient participants were administered measures of cognitive function. Where tests of cognitive function suitable for individuals seven years of age and over (i.e. WISC-III and WAIS-III) were used, the researcher and patient participant were situated in a reasonably private part of the participant's home to maximise participant concentration and confidentiality. This procedure was also adopted when administering questionnaires to patient participants. Often this was done with the door closed or ajar. All of these procedures were tape-recorded for health and safety and reference purposes. At least one parent was present during the cognitive testing of patient participants aged under-seven years. Patients aged under-seven years were not administered questionnaires.

The methodology for the survey, administering of questionnaires, and patient participant cognitive testing was therefore in four parts:

1. MPS Health Assessment and Medical History Questionnaires sent to parents *by post* prior to the researcher's visit.
2. Face-to-face survey with mother.
3. Patient participant questionnaires/cognitive testing.
4. Telephone administration of questionnaires to father.

Tables 4-5 to 4-9 below detail the measures completed by patient (child), parent (mother), and parent (father) participants and their relevant sub-scales. Detailed descriptions of each of the measures plus rationales for their use are provided later in this chapter.

**Table 4-5 Measures Completed by Both Parent (Mother and Father) Participants (where relevant) prior to the Researcher's Visit**

Number of Participants	Measures Completed
44	MPS I Health Assessment Questionnaire – Part One: Child's Physical Functioning
44	Patient Medical History Questionnaire

**Table 4-6 Measures Completed by Patient (Child) Participants**

N	Measures completed	Sub-Scales
22	Behaviour Assessment System for Children Aged 8-11/12+ Years: Self-Report of Personality (Reynolds and Kamphaus, 1998)	Emotional Symptoms Index School Maladjustment Clinical Maladjustment Personal Adjustment
22	Self-Image Profiles for Children aged 7-11/12+ years (Butler, 2001)	Positive Self-Image Negative Self-Image Self-Esteem
23	Mental Development Scales (0-8 years) (Griffiths, 1971)	Locomotor Personal-Social Hearing and Speech Eye & Hand Co-Ordination Performance
16	Wechsler Intelligence Scale for Children – 3 <sup>rd</sup> Ed (6-16 years) (Wechsler, 1992)	Total IQ Verbal Scale Performance Scale
5	Wechsler Adult Intelligence Scale – 3 <sup>rd</sup> Ed (17+ years) (Wechsler, 1998)	Total IQ Verbal Scale Performance Scale

**Table 4-7 Measures Completed by Parent (Mothers) Participants as part of the Parent Survey: Proxy Ratings for Child Health and Psychosocial Outcomes**

N	Measures Completed	Sub-Scales
44	Child Health Questionnaire (Parent Form) (Landgraf, Abetz, and Ware, 1999)	Physical Functioning Limitation of Social Role General Health Pain and Discomfort Impact on Family Limited by Emotional Difficulties Time Impact on Parent Emotional Impact on Parent Child's Self-Esteem Child's Mental Health Child's Behaviour Family Cohesion Change in Health Status
44	Vineland Adaptive Behaviour Scale (Sparrow, Ball, and Cicchetti, 1984)	Communication Daily Living Skills Socialisation Motor Skills
44	MPS I Health Assessment Questionnaire – Part Two: Level of Assistance Required by Child with Activities of Daily Living	n/a
39	Behaviour Assessment System for Children: Parenting Rating Scale for Children aged 2½ years and over (Reynold and Kamphaus, 1998)	Externalising Problems Internalising Problems Behavioural Symptom Index Adaptive Behaviour
5	The Toddler Behaviour Assessment Questionnaire for children aged under-2½ years (Goldsmith, 1996)	Activity Level Expression of Pleasure Social Fearfulness Interest/Persistence Anger Proneness
44	Child Socialisation Scale (measure developed for this study)	Details of individual factors provided in the Measures section of this chapter



**Table 4-8 Measures Completed by Parent (Mothers) Participants as part of the Parent Survey: Parent Measures**

N	Measures Completed	Sub-Scales
44	Parenting Stress Index (Abidin, 1995)	Parent Distress Parent-Child Dysfunctional Interaction Difficult Child
44	Sense of Coherence (Antonovsky, 1979)	Comprehensibility Manageability Meaningfulness
44	General Health Questionnaire (Goldberg and Williams, 1988)	Somatic Symptoms Anxiety and Insomnia Social Dysfunction Severe Depression
44	The Child Abuse Potential Inventory (Milner, 1986)	Distress Rigidity
44	Family Environment Scale (Moos and Moos, 2002)	<i>Relationship:</i> Family Cohesion; Expressiveness; Conflict. <i>Personal Growth:</i> Independence; Achievement Orientation; Intellectual-Cultural orientation; Active-Recreational Orientation; Moral-Religious Emphasis. <i>System Maintenance:</i> Organisation; Control
44	Social Support Questionnaire (measure developed for this study)	Details of individual factors provided in the Measures section of this chapter
44	Family Discord Questionnaire (measure developed for this study)	Details of individual factors provided in the Measures section of this chapter
44	Parent Anxiety about Child Welfare and Perceptions of Offspring Risk (measure developed for this study)	Details of individual factors provided in the Measures section of this chapter
44	Parent Expectations of Child (measure developed for this study)	Details of individual factors provided in the Measures section of this chapter
44	Beliefs and Attitudes (measure designed for this study)	n/a
44	BMT perceptions (measure developed for this study)	Details of individual factors provided in the Measures section of this chapter

**Table 4-9 Measures Completed by Parent (Father) Participants**

N	Measures Completed	Subscales
36	Parenting Stress Index (Abidin, 1995)	Parent Distress Parent-Child Dysfunctional Interaction Difficult Child
36	Sense of Coherence (Antonovsky, 1979)	Comprehensibility Manageability Meaningfulness
36	MPS I Health Assessment Questionnaire – Part Two: Level of Assistance Required by Child with Activities of Daily Living	n/a

### **4.3. Ethical Considerations**

Families were assured of confidentiality and anonymity. The Parent and Patient Information Sheets also informed participants that they could withdraw from the study at any time and that they were not obliged to answer any questions they were not comfortable with. Written informed consent was obtained from all parent participants on behalf of themselves and their children aged under-18 years. Adult patients were asked to give their own consent. Where they were unable to give consent due to learning disability, parents were asked to do so on their behalf. All participants, including child and adult patients, were provided with information sheets, which were appropriately written for children and adult MPS patients. Parents were encouraged to discuss the research with their children before deciding to take part. When the researcher visited, the aims, purpose, and procedure of the research were verbally given to all patient and parent participants. All participants were given the opportunity to ask questions and contact details for the researcher and advocacy support workers at the MPS Society were provided should any post-survey distress be experienced. Confidentiality and the right to withdraw were assured again at this time.

The Written Informed Consent form stated that participants had read and understood the aims and procedure of the study and that they were willing to take part. It also stated that participants gave permission for their child's (or their own in the case of adult participants) GP and bone marrow consultant to be informed of their

participation in the research (See Appendices E and F). Although patient participants aged 18 years and over were asked to give consent, a parent was asked to be present and involved in this process.

Participants were not paid to take part in this study. No identifying information was written on the survey schedule or audio cassettes. Survey schedules were coded with a personal identification number and all coding information was kept separately under lock and key.

As previously mentioned, the Eastern Multi-Research Ethics Committee granted ethical approval for this research, and after some modification at the request of the REC considered all of the written materials provided to the families and methods of gaining consent to be appropriate for children and adults with learning disability.

#### **4.4. Sample Size Considerations**

Since this is the first quantitative study conducted in this area there are no existing parameters from which to statistically calculate the necessary sample size. Additionally, this exercise is complicated by the limitations imposed by the overall number of individuals who comprise the research population. To date there are only 49 children in Britain with this disorder, thus any attempt at calculating sample size is largely academic. Since the logical method for analysing the data in light of the research hypothesis would be to use path analysis, a developed form of multiple regression, the criteria regarding the number of participants for this statistical procedure provides the basis for this calculation. However, due to the small sample size, it is inappropriate to attempt to obtain goodness of fit for the path. Therefore, a series of multiple regression analyses will be employed to draw up a theoretical path.

#### **4.5. Data Processing and Analysis**

1. Pearson product-moment correlations were computed between all variables to explore relationships. This helped determine the variables that were entered into the regression models.

2. A series of hierarchical multiple regression analyses were employed to investigate the moderating effects of resistance factors - intrapersonal, socio-ecological, and stress processing - on the effects of illness-related and psychosocial stressors on parent and child adjustment. Data were entered into the analyses in theoretical groups as informed by the literature reviewed and correlational analyses conducted as described in point 1 above.
3. Discrepancy variables were created and entered into the multiple regression analyses to determine whether divergence or a shared view of parenting stress, coping, and child care needs impacted upon parent and child adjustment as a social-ecological resistance factor.

## **4.6. Measures**

As previously stated, data were collected in four phases. Firstly through postal questionnaires which were sent to mothers and fathers jointly (where applicable) prior to the researcher's visit; Secondly via a survey, which was administered by the researcher to mothers only. Thirdly, questionnaires were administered to patient participants aged seven years and over and all patient participants administered tests of cognitive function. In the fourth and final phase, fathers were administered questionnaires over the telephone.

The survey was administered to mothers face-to-face and was researcher-led. This allowed for the potential misinterpretation or misunderstanding of questions, and for potential literacy difficulties, as the researcher not only verbally delivered the questions but could clarify any misunderstandings mothers had regarding the questions. The survey coding was pre-categorised, but in addition, actual responses or quotes from participants were recorded where these were apposite, or equally where unexpected or unusual views were expressed. To assist participants in selecting their responses, laminated answer cards were provided for some of the measures where specific answer options or scaled answer options were needed. The survey comprised a number of published measures, together with demographic and biographical questions. Some new measures were also designed specifically for this population. The main areas covered in the parent survey are detailed below, followed by descriptions and psychometric properties of each of the published measures. New

measures are also described and information regarding piloting and internal reliability given. Demographic and biographical questions can be found in Appendix L, published measures in Appendix N and O, and new measures in Appendix M.

#### **4.6.1. Design and Piloting of New Measures**

While the psychological impact of chronic illness on children and families may be the manifestation of processes common to all chronic illness (Stein and Jessop, 1982a), there are many factors that are particular to specific medical conditions that have been found to impact on families, as discussed in the literature review (e.g. Brewer et al., 2008; Suls, 1982; Williamson, 1999). This can particularly be the case when the condition is rare and genetically transmitted. It was therefore deemed important to further examine a number of issues highlighted in the qualitative phase of this study that related in particular to the adjustment of this patient group and which were not covered by existing instruments. A number of measures were therefore designed to tap into psychological constructs, such as uncertainty, perceptions of child vulnerability, and perceptions of support and understanding, by making reference to or inferences about issues that were specific to the experiences of this population, such as the bone marrow transplant, the terminal nature of the untreated condition, functional independence, and the progressive nature of bone and joint disease. It is hoped that the examination of constructs that are particular to this patient group and their families will add to knowledge about specific risk and resistance factors, which in turn may aid the development of specialist support services. The new measures were devised as follows:

1. Child Socialisation
2. Parent Anxiety about Child Welfare and Perceptions of Offspring Risk
3. Parent Expectations of Child
4. Social Support
5. Family Discord
6. Beliefs and Attitudes
7. Satisfaction with Schooling
8. Parental Perceptions of BMT experience

The measures were designed by the author of the present study. The items and sequencing of items on each of the measures were informed by discussions with experts in the field of MPS disorders, and by the qualitative interviews with MPS families in the first phase of this project. All new questionnaires were piloted with parents of children affected by MPS diseases similar to MPS IH post-BMT. These families were not included in the study however. Three of the new questionnaires: The Child Socialisation Questionnaire, Parent Anxiety/Risk Perceptions Questionnaire, and Social Support Questionnaire were piloted with 49 mothers of children with other MPS Diseases, including MPS IV Morquio Disease, MPS I Hurler-Scheie Disease, and MPS II Hunter Disease. They were also piloted with 46 parents of healthy children. The remaining two questionnaires: Family Disagreements and Parent Expectations of Child were piloted with the 49 families of children with other MPS diseases only, as the line of questioning was not relevant to parents of healthy children, as they referred specifically to the MPS condition and the BMT. With the inclusion of the parents of children affected by MPS IH post-BMT who participated in the second phase of this study, a total of 139 families completed some or all of the above new measures. The timing of this piloting phase can be seen in the study flow chart in Appendix A.

The methodology for this piloting process consisted of questionnaires being sent out with a covering letter and information sheet, explaining the purpose and procedure of this process, to all parents of children aged 18 years and under affected by the above MPS disorders. They were all recruited via the MPS Society in the UK. The questionnaires were received anonymously and consent to participate was obtained from the parents. Parents of children not affected by chronic illness or disability were

recruited via local schools. Questionnaires were distributed anonymously to parents via the schools' Head Teachers. They were accompanied by a covering letter and information sheet as above. All parents were assured of confidentiality and anonymity.

Including data collected from parents of children affected by MPS IH post-BMT for the second phase of this study, the items in each of these measures were subjected to Factor Analysis. For each Factor Analysis the Principle Components method was used, with varimax rotation, revealing factors with eigenvalues higher than 1.0. An inclusion criterion included that factor loadings had to be 0.4 or above. This was then followed by tests of Internal Consistency using Cronbach's Alpha. Details of each of these procedures are included in the measure descriptions below and the Factor Analysis tables can be found in Appendix P.

#### **4.6.2. Measures Completed by Both Parent (Mother and Father) Participants Prior to the Researcher's Visit**

##### MPS I Health Assessment Questionnaire – Part One

The MPS I Health Assessment Questionnaire (MPS I HAQ) was administered to parents as a measure of patient physical functioning. Physical functioning has been highlighted as a crucial risk factor in both patient and parent adjustment to living with chronic illness and has been linked to adjustment disorders (e.g. Witt et al., 2003). The MPS I HAQ is the only MPS-specific quality of life instrument that was in development by Genzyme Corporation at the time of this research and was yet to be validated. Its use here was permitted with the intention of assisting with that validation process. It was noted however that other validated measures of physical functioning such as the SF-36 (Ware and Sherbourne, 1992), FS II(R) (Stein and Jessop, 1990), and WHO-QOL would also have been appropriate. However, the support of the validation of an MPS-specific measure of physical functioning was decided upon over the use of these measures.

The MPS I HAQ is in two main parts. The first part questions the ability of MPS patients to perform various activities of daily living, including eating/drinking,

dressings, toileting, bathing, grooming, walking, gross motor skills, and fine motor skills. Each item asks the parent or patient to rate the level of difficulty they experience performing each activity. Responses are given using an anchored scale of 0-10 ('not difficult at all' to 'extremely difficult'). The use and frequency of use of mobility aids is also explored. This measure has enabled the formulation of an accurate picture of physical functioning, including essential self-care activities, the demise of which can impact on quality of life. Part One of this measure can be found in Appendix Li. The second part of the scale explores the extent to which patients require assistance with a number of activities. Again these include activities such as eating, bathing, dressing, transfers, and indoor and outdoor locomotion. These items are rated using parameters of assistance from 'independent' to 'complete assistance'. Part Two is included in the survey schedule, copied in Appendix K. Alpha coefficients for each sub-scale on this measure and the total scores for parts one and two were in the .80s and .90s for this study sample (N=43). Thus, all subscales and total scores on this measure achieve a satisfactory level of internal reliability (Nunnally, 1967).

The first part of this measure was sent to the families prior to the researcher's visit. The second part formed part of the survey administered to mothers. It was also administered to fathers by telephone. The purpose of this was to ascertain whether parents perceived their child's care needs differently from one another. As reflected in the findings of the qualitative phase of the study and the literature reviewed, one parent often takes responsibility for the day-to-day care of the child and would thus have more knowledge of their support and care needs. As also reflected mothers can become ensconced in their caring role and over-protect their child, thus perceiving them to need more support than they perhaps do. Differential concepts of the child's care needs between parents in this study could therefore reflect the different ways in which the child's illness impacts on each parent both physically and psychologically and their differing commitment to the caring role. This could have implications for the kind of support parents can give one another, for mother's psychological well-being, and for parenting stress and coping. These factors, in turn, could have implications for child psychosocial development.



## Patient Medical History

Medical events and conditions are an unfortunate part of living with MPS IH post-BMT. Most of the children have or will at some time in their lives have significant orthopaedic surgery to correct spinal curvatures, genu valgum (knock-knees), hip dysplasias, and carpal tunnel syndrome. Some of the children have eating difficulties post-BMT, some have experienced on-going respiratory difficulties and lung infections due to graft versus host disease, and some experience cardiovascular difficulties. They therefore spend significant amounts of time in hospital throughout their lives. Furthermore, since medical and orthopaedic outcomes of BMT are uncertain, this group of children are regularly monitored by their MPS Consultants (Paediatricians). Regular hospital visits; x-rays, and blood-taking therefore form part of their normal lives. Data from the qualitative phase of this study illustrated how some of the children have been significantly emotionally affected following bouts of illness, infections, and surgery and how this impacts on parents' well-being and their ability to cope. It was therefore felt important to obtain a record of medical attention received in the children's lifetime to see if these experiences impact on both parent and child adjustment.

A set of questions was therefore devised which aimed to record the total number of times the child has visited the GP, local consultant, and specialist MPS consultant in the previous six months; how many times they had visited both their local hospital and specialist MPS hospital for routine checks in the last 12 months, and how many nights in the child's lifetime they had spent in hospital and/or ICU. It also asked parents to record the number of times the child had visited A&E in their lifetime and the number of surgical procedures they had undergone, orthopaedic and otherwise; whether they had any ongoing medical problems and were on medication, whether they were visually and/or hearing impaired and required aids, whether they suffered from any mental health disorders and took medication, whether they had experienced early puberty (if of the appropriate age) and whether or not they had experienced a serious adverse events in their lifetime. Examples of serious adverse events were given to assist parents in deciding whether or not their child had experienced any. The purpose of asking these questions was to ascertain parents' perceptions of their child's health care needs. Since this information could not be verified and was subject to

parents' interpretation, it could not be an objective or accurate record of the child's medical history. It could reflect however, parents' perceptions of child vulnerability and their concern for this child in terms of their health, which was demonstrated in the findings of the qualitative phase and in the literature reviewed to potentially impact upon parent well-being and coping. It could also serve as a proxy for disease severity. A copy of the patient medical history questionnaire can be found in Appendix Lii.

#### **4.6.3. Psychosocial and Developmental Outcome Measures Completed by Patient (Child) Participants**

##### Self-Image Profiles (Butler, 2001)

The self-image profiles (SIP) are brief self-report measures that explore the individual's perception of self. They were administered here to patient participants. There are two forms; the SIP-C for children aged 7-11 years and the SIP-A for adolescents aged 12+ years. The SIP provides a visual display of Self Image, enabling the child/young person to illustrate the way in which they see themselves as they complete it. All items are self-descriptions comprising words or statements that have been generated by children and adolescents. The words used in each item are therefore meaningful to the population for which this scale has been designed. With the simple and age-appropriate wording, and only 25 items, these profiles were felt to be accessible to the younger individuals in the present sample, and to young people with mild to moderate learning difficulties. Each item uses a 6-item Likert-type scale, with which the respondent rates *how much they are like* the statement or word presented. The measure comprises twelve positive items, twelve negative items, and one neutral item. They provide two total self-image scores: positive self-image and negative self-image. They also provide a measure of self-esteem. The Self-Image Profiles were verbally administered by the researcher to all patients aged seven years of age and older, who were able to complete it (n = 22). Due to the nature of the measure and the level of understanding required, it was not designed for use with children under 7 years of age. Thus, children aged under-7 years (n = 19) and those with more severe learning disabilities (n = 3) were not administered this measure. Of the children that completed this measure 12 were aged 7-11 years and 10 were aged 12 years and over. This measure can be found in Appendix Oi.

The Behaviour Assessment System for Children (BASC) Self-Report of Personality (Reynolds and Kamphaus, 1998)

A self-report of personality has been designed for ages 8-18 (8-11; 12-18), which comprises 152-186 items relating to a child/young person's thoughts and feelings through a series of true/false questions. These items are grouped in a number of subscales, such as anxiety, attitude to school, interpersonal relationships, depression, self-esteem, and social stress. All items are statements, such as "my classmates don't like me", which require a true/false response. This measure was given verbally by the researcher to all patient participants aged 8 years and over, unless difficulty was experienced completing this measure. Eighteen patient participants completed this measure, 10 aged 8-11 years and 8 aged 12 years and over. Nineteen patient participants did not participate because they were too young to meet the age criteria for the measure, and 7 did not participate due to learning disability. In some of these cases, only the Butler profiles were administered. Although there is a self-esteem component to this measure, it has a far wider scope than the Butler profiles, which facilitated the exploration of children and young people's feelings about a variety of aspects of their lives. It also allowed for the potential identification of adjustment disorders. With regard to scoring, this measure provides an overall composite score, the Emotional Symptoms Index. However, it also provides composite scores of School Maladjustment, Clinical Maladjustment, and Personal Adjustment. It has both positive and negative items, and may be interpreted in relation to National norms. Internal consistencies of the scales are high, averaging about .80 for each gender at both age levels. This measure can be found in Appendix Oii.

Mental Development Scales (0-8 years) (Griffiths, R., 1971)

The Mental Development Scales were administered to patient participants as a measure of cognitive functioning. Cognitive development has been used as a measure of functional (in) dependence as a risk factor in the adjustment of chronically ill children (e.g. Casey et al., 2000). The Scales measure patterns of development that are significant for intelligence or mental development in babies and young children from birth to 8 years of age. The Scales are split into two age groups, 0-2 and 2-8 years,

and different apparatus and tests are required for the different groups. While some of the items are common to both scales, there are 27 tests for the younger age group, and 22 for the older age group. The subscales include locomotor, personal-social, hearing and speech, eye and hand co-ordination, and performance. However, an overall GQ is calculated from these scales. In relation to the present study, the Scales were completed for the most part by children aged 6 years and under, with the exception of one adult participant, two 10-year olds, and a 13-year old. This was due to the individuals' learning disability. The decision for older patient participants to be administered this test due to learning disability was made only after the scale appropriate for their age was first administered and the first few items failed. Consent for these individuals to participate in this type of testing was sought from the attending parent, usually the mother. A.R.I.C.D. approve of the Griffiths Mental Development Scales being used as a measure of developmental functioning in older children with learning disabilities (personal communication with the Honorary Secretary of A.R.I.C.D. January 2009). The scales are supplied only to professionals who have undergone training in their use and application. The author underwent such training in 2004 through ARICD. This measure can be found in Appendix Oiii.

The Wechsler Intelligence Scale for Children – 3<sup>rd</sup> Edition (WISC-III UK) (6-16 years) (Wechsler, D., 1992)

The WISC-III is a test of intellectual ability and was also administered to patient participants as a measure of cognitive function. It is a collection of subtests, which are divided into two scales – Verbal and Performance. It is designed for use with children between the ages of 6 and 16 inclusive, and is designed to be completed without reading or writing. For the purpose of the present study however, some individuals aged over-16 years completed this measure, as a result of their learning disability. As with the Griffiths Mental Development Scales, the test appropriate for the patient's age was first administered and on failure to complete the first few items, the decision was made to use the test suitable for younger individuals. The Verbal Scale tests use language-based items, while the Performance Scales use visual-motor items that are less dependent on language. Scale-specific IQs are produced, as is a Full Scale IQ. This measure can be found in Appendix Oiv.

Wechsler Adult Intelligence Scale – 3<sup>rd</sup> Edition (WAIS-III UK) (17 years plus)  
(Wechsler, D. 1998)

The WAIS-III is a test of intellectual ability designed for individuals aged 17 years and above. It has 14 subtests, which produce a Full Scale IQ, Verbal and Performance IQs, and other indexes, including Verbal Comprehension, Perceptual Organisation, Working Memory, and Processing Speed. This measure was administered to patient participants and can be found in Appendix Ov.

**4.6.4. Measures Completed by Mothers as Part of the Parent Survey**

**4.6.4.1. Demographic Measures**

Demographic information was gathered as follows:

Mother's relationship to child; biological parenthood of mother's partner; number of children in the family, family composition, and child's birth order. Parents' ages, ethnicity, and first language were also obtained. As was parents' education, employment, and income status. The purpose of this was to gather descriptive demographic information about the families for interest purposes, but also to explore utilitarian family factors and their relationship to parent and child adjustment (Wallander et al., 1989). These questions can be found in the survey schedule in Appendix K.

**4.6.4.2. Proxy Ratings of Child Health and Psychosocial Outcome Measures**

Patient MPS and Bone Marrow Transplant details

Historical BMT and MPS-specific information was gathered as follows:

Child's gender, child's age at the time of the study, length of time since the last transplantation, age at transplant (first and second), donor identification information, type of transplant, and MPS I genotype (genetic code of inherited maternal and

paternal faulty genes). The purpose of gathering this information was to allow for the exploration of relationships between variables such as child age and gender or length of time caring and parent and patient adjustment. It also allowed for developmental outcomes to be examined in relation to the length of time since the transplant as opposed to age, as later treatment can have implications for cognitive and adaptive functioning. Information regarding BMT type and MPS genotype allowed for the verification of patient homogeneity in these respects to rule out the possibility of adjustment outcomes being due to treatment type or disease severity rather than the risk and resistance factors being explored. MPS genotype illustrates the severity of the condition and allowed the researcher to verify whether or not all patient participants were affected by the most severe form of MPS I, Hurler disease. For the most part parents did not know their child's MPS genotype. It was therefore gathered from the patients' respective MPS-specialist hospitals, thus medically verified.

Further historical bone marrow transplant information was gathered which pertained to whether or not the child experienced complications during the procedure, whether they contracted graft versus host disease (GvHD), and whether or not they continued to experience associated problems. Mothers were asked to rate the seriousness of these problems as mild, moderate, or serious. The ratings were therefore subjective and subject to mothers' interpretations. The purpose of this line of questioning was to establish mothers' perceptions of their child's health status in relation to the BMT. In cases where the child was regarded as experiencing serious complications during the BMT, contracting GvHD, or continuing to experience problems post-BMT, it may indicate perceptions of child vulnerability, which may be useful to the investigation of both parent and child adjustment outcomes.

The total number of MPS pregnancies that resulted in termination and the number of MPS pregnancies that resulted in a live birth was also recorded. This allowed for further description of family demographics. These questions can be found in the survey schedule in Appendix K.

#### MPS I Health Assessment Questionnaire – Part Two

See point 4.6.2 on pages 190-191 for details.

The Child Health Questionnaire (CHQ – Parent Form) Landgraf, Abetz, and Ware, 1999)

The CHQ is a family of generic quality of life instruments designed for children 5 years of age and older. Some scales on the parent form can be used in younger children, though the scale has not been validated for use with under-5s. It is completed by parents and was administered to mothers in the present study. The parent form has 50 items, which employs a Likert-type scale whereby higher scores indicate a more positive health status. Scores can be analysed separately (CHQ profile scores) or combined to derive an overall physical or psychosocial score (CHQ summary scores). The parent version used for the present study has been standardised in the UK. The scale has 14 subscales as follows: Parent assessments of the child's physical functioning, the degree to which the child is limited in their role and social activities by their physical limitations (including participation in school work, activities with friends, physical activities, and self-care activities), the child's general health, the child's bodily pain and discomfort, the impact the child's health has on family activities, the degree to which the child is limited in their role and social activities by emotional difficulties, the time impact the child's illness has on the parent, the emotional impact the child's illness has on the parent, the child's self-esteem, the child's mental health, the child's behaviour, family cohesion, and whether or not the child's health has changed in the last year. These sub-scales were applied as illness-related risk factors and as stress processing factors in the investigation of predictors of parent and child adjustment outcomes.

In 6 out of 10 clinical samples 86% or more of the alpha coefficients met or exceeded .70, indicating acceptable internal consistency (Landgraf et al.,1999). For the present study sample the alpha coefficients for all subscales exceeded .70, with the exception of Mental Health (.52) and General Health Perceptions (.53). This measure can be found in Appendix Ni.

The Vineland Adaptive Behaviour Scale (VABS) (Sparrow, Balla, and Cicchetti, 1984)

The findings of the qualitative phase of this study illustrated patient functional difficulties, particularly in the areas of communication and socialisation. The VABS is an interview-based measure, which evaluates children's functional difficulties in these areas, as well as in daily living skills. All of which may be related to patient quality of life. This scale is administered in interview format by a researcher to the parent or caretaker, and has age-relevant questions spanning infancy to early adulthood. It can also be given when a child is quite debilitated. It was administered to mothers in the present study. This assessment provides critical data for the evaluation of a wide range of disabilities, including learning disability, developmental delays, functional skills impairment, and speech and language impairment.

The measure explores four major domains of adaptive behaviours, which themselves are broken down into further sub-domains, as follows:

<u>Domain</u>	<u>Sub-Domain</u>
Communication:	Receptive; Expressive; Written.
Daily Living Skills:	Personal; Domestic; Community.
Socialisation:	Interpersonal relationships; Play and leisure time; Coping skills
Motor Skills:	Gross; fine

This measure facilitates the exploration of differences in adaptive/maladaptive behaviour across age groups and gender, and in relation to disease-related, individual, social, and family factors. For the present study, it was administered to the mothers of all patient participants. While, like the interview questions in the qualitative phase of this study, this measure can only glean mothers' perceptions of their children's behaviour and abilities, it allows for a more comprehensive and systematic exploration of adaptive functioning post-BMT and allows for comparisons to be made within the MPS group and in relation to norms. Separate independent measures of patient development and cognitive functioning were also administered by the researcher to allow for these limitations.



This measure has previously been utilised to measure functional (in)dependence as a risk factor in the adjustment of chronically ill children (e.g. Casey et al., 2000), and it has been shown to have good psychometric properties, with good split-half, test-retest, and inter-rater reliability as well as adequate construct, content, and criterion-related validity (Sparrow et al., 1984). Internal consistency on the adaptive and maladaptive behaviour composites range from .77 to .98. Using Cronbach's Alpha, the internal consistency for the Adaptive Behaviour Composite for the present study sample was .92 with motor skills included and .90 with motor skills excluded. Thus, confirming that this measure achieves a satisfactory level of internal reliability. This measure can be found in Appendix Niv.

The Behaviour Assessment System for Children (BASC) Parenting Rating Scale (PRS-C) (Reynolds and Kamphaus, 1998)

The BASC was developed from the belief that all children have strengths that should be recognised and used to help remedy behavioural and emotional problems. The BASC therefore includes both positive (adaptive) and negative (maladaptive) aspects of the child. The Parent Rating Scale has forms at three age levels: preschool (2 ½ - 5 years), child (6–11 years), and adolescent (12-18 years). For the purpose of the present study, the 12-18 year version was administered to the mothers of all patient participants twelve years of age and over. This was done for consistency purposes. The Parent Rating Scale contains 126-138 items and measures adaptive skills, as well as externalising problems and internalising problems. The forms contain descriptions of behaviours that the respondent rates on a four-point scale of frequency, ranging from *Never* to *Almost Always*. Internal consistency reliabilities of the composite scores are in the middle .80s to .90s at all three age levels. For the present study sample however, internal consistency reliabilities scored as follows:

	<u>2-5 years</u>	<u>6-11 years</u>	<u>12-18 years</u>
Externalising	.701	.782	.646
Internalising	.599	.790	.515
BSI	.776	.851	.744
Adaptive Behaviour	.547	.693	.904

Some composites therefore have low internal reliability scores with this study sample. However, the BASC has well-established internal consistency, reliability, and validity (Doyle, Ostrander, Skare, Crosby, & August, 1997; Reynolds & Kamphaus, 1992), and is widely used for the purpose of diagnostic assessment. (Reynolds & Kamphaus, 2004). Research also supports the validity of the Parent Rating Scale and Teacher Rating Scale for the assessment and identification of children presenting with attention-deficit/hyperactivity disorder (Doyle et al., 1997).

Although, as with the Vineland Adaptive Scales, this is a measure of mothers' perceptions of child behaviour, it permits a systematic examination of relationships between clinical manifestations of personality, such as conduct disorders, anxiety, depression, and withdrawal, and adaptive personality traits such as sociability and leadership skills, which the qualitative phase interview did not facilitate. It also allows for the exploration of relationships between risk and resistance factors and behavioural outcomes using quantitative methods. In the qualitative phase of this study some of the children were described by their parents as being withdrawn, inhibited, and socially anxious, and in one case to show obsessions and compulsions. Other children were described as being confident, outgoing, and sociable. These findings warranted further investigation so that these differences could be explored in relation to disease-related and psychosocial factors. This measure can be found in Appendix Niii.

#### The Toddler Behaviour Assessment Questionnaire (TBAQ) (Goldsmith, 1996)

This measure comprises 6 scales and 108 items, and was administered to mothers as part of the survey schedule. It is designed to examine temperament-related behaviour in 16-36 month old children. The scales include activity level, anger, fear, pleasure, and interest. This was felt to be a useful measure of temperament at this crucial age as the children affected by MPS IH in the present study sample had recently received their bone marrow transplant and were not long out of isolation. They were commonly reported by their parents in the qualitative phase of this study as showing signs of distress, fear of novel situations, and withdrawal in the months following BMT.

Furthermore, no research has explored infant recipients of BMT affected by MPS IH and their responses following treatment. This instrument therefore enabled the systematic investigation of these behaviours among this age group, which the BASC could not due to its age parameter cut-off of 2 ½ years. The TBAQ was administered to the mothers of infants aged under 2 ½ years, which constituted 11% of the study sample population. While this was a small sub-sample size, it was felt important to explore the children's behaviours in detail so that future research of this nature with this patient group can be informed.

This questionnaire has internal consistency reliability typically exceeding .80 for each scale. Evidence for convergent validity with other temperament questionnaires has also been obtained. However, since there are many items on this measure and only five participants in this age group in this study sample, tests of internal consistency proved impossible to complete for all subscales. For those subscales that had enough variance on each item, internal reliability scores using Cronbach's Alpha are as follows:

Activity Level = .891

Expression of Pleasure = .697

Interest/Persistence = .885

Anger Proneness = .677

This measure can be found in Appendix Nii.

### Child Socialisation

This measure was developed for use in this study. It was designed as an outcome measure of patient psychosocial adjustment but also for use as an intrapersonal resistance factor for patients' psychosocial adjustment in other areas. It was administered to mothers as part of the survey. The data from the qualitative phase of this study illustrated parents' concern for their children in terms of their social competence. It also highlighted the degree to which parents allowed their children to participate in social activities away from the home, and their concern for their welfare if they were to participate in activities without their supervision. Many spoke of their

children's lack of friendships and social inhibition. However, this was not the case for all, and some parents described their children as sociable and outgoing, and for their participation in social activities to be encouraged. The literature reviewed also highlighted the protective nature of friendships and social support to child adjustment (e.g. LaGreca et al., 1995; LaGreca and Thompson, 1998), and illuminated social assertion as an intrapersonal variable that has been related to child and adolescent adjustment (e.g. Livneh et al., 2004). How much patients participated in social activities as facilitated by their parents was therefore considered to warrant further investigation. A set of five questions was created to explore the frequency with which parents enable their MPS children to participate in social activities outside of nursery, school, or college. Two versions of the measure were created in order to account for age-appropriate activities: Under-12 years and 12 years and over. However, the items across each age group are comparable. Each item asks the parent to state how often their child participates in various social activities using the following scale and key:

1	2	3	4	5
Never	Rarely	Sometimes	Quite often	Frequently

*Key*

*Frequently: At least once a week*

*Quite often: Once to twice a month*

*Sometimes: Every other month*

*Rarely: 2-3 times a year*

*Never: Never*

This is with the exception of item three in the 12+ version, which asks whether the child has ever gone on holiday without his or her parents. The following answering scale was used for this item:

1	2	3	4	5
Never	Once or Twice	Has been a few times	Every other year	Goes regularly (1-2 times per year)

This measure was completed by all three pilot groups: MPS "other" MPS IH and Healthy Child. The Child Socialisation measure can be found in Appendix Mi.

The Principle Components Analysis for the under-12s version of this measure revealed that the scale can be broken down into two factors. Factor 1 corresponds with the dimension of General Socialisation with Peers, and Factor 2 with Organised Activities. The percentage of variance explained by each factor was calculated as: Factor 1 = 49.1% and Factor 2 = 34.4%. This explained 83 percent of the total variance. The Factor Analysis table for this version of the measure can be found in Appendix Pi. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: General Socialisation with Peers  $\alpha = .928$ , and Organised Activities  $\alpha = .693$ . Thus indicating that Factor 1 achieves a satisfactory level of internal reliability, while Factor 2 achieves a level of reliability that is borderline (Nunnally, 1967).

The Principle Components Analysis for the 12 years and over version of this measure revealed that the scale has one factor with eigen values higher than 1.0. As the solution could not be rotated, the component matrix is shown in the Factor Analysis table presented in Appendix Mii. Factor 1 corresponds with the dimension of General Socialisation with Peers including organised activities. This singular factor explains 60 percent of the total variance. Computation of the internal consistency for this one factor using Cronbach's Alpha revealed the following score: General Socialisation with Peers including Organised Activities  $\alpha = .829$ . Thus indicating that this measure achieves a satisfactory level of internal reliability (Nunnally, 1967).

#### **4.6.4.3. Parent Outcome Measures**

##### Parenting Stress Index – Short Form (Abidin, 1995)

The Parenting Stress Index (PSI) was designed to produce a measure of the extent to which stress is experienced in the parent-child system. It is a screening and diagnostic instrument that was developed using the concept that the amount of parenting stress experienced is dependent upon child characteristics, parent characteristics, and situations that are directly related to the role of being a parent. There is a short form for this instrument, which consists of 36 items, and comprises three factors, which capture the primary components of the parent-child system – Parent Distress, Parent-Child Dysfunctional Interaction, and Difficult Child. It was given to mothers at part of

the survey and to fathers by telephone in the same form. The Life Stress measure from the original scale was also administered to mothers as part of the survey to account for any recent life events encountered by the family. This measure is useful to the present study, as it helps to identify potential dysfunctional parent-child systems. It can tap into family functioning and parenting skills, parental self-efficacy, and the spousal relationship. When given to both parents, this measure also allows the comparison of spousal experiences of parenting stress. Parenting stress was used here as an outcome measure of parent adjustment. It was also used as a stress processing factor for exploration in relation to other parent and child adjustment outcomes.

Normative data has been collected for a large sample of mothers and a small sample of fathers, from a variety of ethnic backgrounds. The measure also has good psychometric properties. Reliability coefficients for this study sample ranged from 0.76 to 0.90. This measure can be found in Appendix Nix.

#### Sense of Coherence (Antonovsky, 1979)

The Sense of Coherence (SOC) was administered to parents as a measure of coping. It is a theoretic construct, which refers to the internal resources that enable individuals to cope with stressful situations, and maintain psychological and physical health. It is a tri-dimensional construct, which measures an individual's global perception of their life and the world around them. The three components that comprise the measure were derived from the analysis of a number of qualitative interviews and are theorised to determine individuals' perceptions and interpretations of external events as follows:

- *Comprehensibility*: The social world is interpreted as rational, understandable, structured, ordered, consistent, and predictable. Comprehensibility is a dimension that refers to the cognitive controllability of one's environment (i.e. individuals with a strong SOC have the ability to define events as less stressful).
- *Manageability*: This denotes the extent to which individuals consider resources to be personally available to them to help them cope adequately with demands or problems (i.e. to be able to mobilise resources in order to deal with stressors).

- *Meaningfulness*: This represents the motivational component and determines whether a situation is appraised as challenging, and whether it is worth making commitments and investments in order to cope.

Parents' perceptions of their child's illness and the situation they were in, the impact it had on them and their ability to cope emerged as themes in the qualitative phase of this study. Coping is therefore an important measure of adjustment which warrants further investigation with the parents of this patient group. The SOC scale is a general measure of an individual's world-view, and can identify internal coping resources. SOC can be seen as a relatively stable (trait) measure. However traumatic events, such as having a terminally ill child, may change a parent's world view, and thus their SOC. The impact that the child's condition might have had on parents' ability to cope can therefore be usefully explored using this measure. The measure as a whole and its sub-scales also provide measures of stress processing (SOC, comprehensibility, meaningfulness) and intrapersonal (manageability) factors, the moderating effects of which can be explored with other parent and child adjustment outcomes. Research has found measures of parental SOC to be related to parent adjustment when they have a chronically ill child (e.g. Olsson & Hwang, 2002). It is a measure that has also proved useful to a number of studies, including those of carer well-being (e.g. Wagenfeld et al, 1994), and those pertaining to parent well-being and child health (e.g. Groholt et al, 2003). It is proposed that this measure may also be a crucial predictor of child psychosocial outcomes (child coping and self-esteem in particular). It has good internal consistency (0.82 – 0.95) (0.85 – 0.93 for this study) and stable test re-test reliability (Antonovsky, 1993).

The SOC was presented to the mother as part of the survey and to the father by telephone. This allowed for the exploration of parental differences in terms of coping. This measure can be found in Appendix Nvi.

#### General Health Questionnaire (GHQ) (Goldberg and Williams, 1988)

The literature reviewed illustrated how women are more likely than men to take responsibility for the day-to-day care of a sick child (e.g. Anderson and Elfert, 1989; Guberman et al., 1992; Parks and Pilisuk, 1991), to simultaneously take care of the

upkeep of the home (Pelchat et al., 2003), deal with illness-related stressors and the routine stressors of everyday life (Goldner, 1985; Thompson et al., 1993), and as a result may be prone to psychological difficulties (Manuel, 2001; Pelchat et al., 1999a; Pelchat et al., 1999b). It was therefore felt important to explore mothers' mental health for the present study, both as an outcome measure of maternal adjustment outcome but also as an intrapersonal resistance factor for exploration with child psychosocial outcomes. Two measures were used to assess mothers' mental health: the GHQ as described here and a subscale of the CAPI as described below.

The GHQ is a screening test designed to detect non-psychotic psychiatric disorders in community settings. It was administered to mothers here as a measure of mother's psychological well-being. The GHQ detects an individual's inability to carry out every day activities, and the manifestation of novel and distressing phenomena. It has established psychometric properties (internal consistency ranging from .77 to .92) and has been widely validated. The original measure is a 60-item questionnaire, which has been scaled down to 30-items and 28-items. The GHQ-28 is used mainly for research purposes, and has four scales: somatic symptoms, anxiety and insomnia, social dysfunction, severe depression. This version is quick to administer and reliable. It was proposed for this research as mother's mental health has been highly related to child emotional well-being in previous research, and this measure in particular has been effectively utilised in studies exploring determinants of child adjustment (e.g. Farmer and Markus, 1986). Internal reliability for this study sample ranged from 0.87 to 0.94. This measure can be found in Appendix Nvii.

#### The Child Abuse Potential Inventory (CAPI) (Milner, 1986)

The CAPI was administered to mothers as an additional measure of their psychological well-being. Previous research has found the CAPI to be an excellent measure of parenting style as well as psychological distress. It is a self-report instrument in which participants are asked to respond 'agree' or 'disagree' to 160 items. There are six scales: Distress, Rigidity, Unhappiness, Problems with child and self, Problems with family, and Problems with others, as well as three validity scales: Lie, Random Response, Inconsistency. For the present study however, only two scales from the inventory were used: Distress and Rigidity. The title of the inventory was not



used in the administration of the scale, nor was the purpose of its use to identify child abuse. The rationale for using only the Distress and Rigidity scales is based on research carried out at the University of Minnesota (Hughes et al., 2003), which found Rigidity in parenting to form part of a parental assets factor, which correlated with maternal intelligence and a home environment that fostered cognitive development. Conversely the Distress factor formed part of a parental distress factor, which was found to correlate with another measure of distress on the Brief Symptom Inventory (Derogatis and Melisaratos, 1983). By administering the CAPI, it may be possible to determine two important dimensions of parental fostering of child development. For the present sample, internal reliability for these two subscales is good: Rigidity 0.77 and Distress 0.96. This measure can be found in Appendix Nviii.

#### **4.6.4.4. Measures Relating to Social-Ecological Factors**

##### Family Environment Scale (Moos and Moos, 2002)

The Family Environment Scale assesses the social climate of all types of families, and evaluates the social environment of the family unit. It is a 90-item scale, with 10 subscale groupings. It can be administered to parents and to children upwards of 11 years of age. However, for the present study it was administered to mothers only. The scales include: Relationship (family cohesion, expressiveness, conflict); Personal growth (independence, achievement orientation, intellectual-cultural orientation, active-recreational orientation, moral-religious emphasis); and System Maintenance (organisation, control). With regard to *relationship* the three subscales explore family commitment and support, the expression of feelings, and the expression of anger and conflict among family members. *Personal Growth* assesses individual family members' assertiveness, self-sufficiency, and autonomy (independence). It assesses the extent to which activities (such as school or work) are cast into achievement-oriented or competitive frameworks (achievement orientation); and measures family members' interest in political, intellectual, and cultural activities (intellectual-cultural orientation). Participation in social and recreational activities is measured (active-recreational orientation); as is the emphasis put on ethical and religious issues and values (moral-religious emphasis). *System Maintenance*: two further dimensions measure the importance of organisation and structure in planning family activities and

responsibilities, as well as the degree to which set rules and procedures are used to run family life (organisation, control). The Family Environment Scale has good test re-test reliability and internal consistency. Normative data are also available from 1,432 'normal' and 788 distressed families.

The findings of the qualitative phase of this study and the literature reviewed demonstrated the importance of family functioning to both parent and child adjustment. It therefore warrants further investigation with this patient group and in relation to parent and child adjustment outcomes. This measure, in particular, has been used to explore family functioning as a social-ecological resistance factor in a number of studies, including those which have employed the disability stress-coping model (e.g. Burlew et al., 2000; Manuel, 2001; Wallander et al., 1989; Wallander and Varni, 1992). This measure can be found in Appendix Nv.

### Social Support

A prominent theme that emerged from the qualitative data illuminated the importance of family relationships and friendships, and how they provided parents with essential support. Conversely, however, the data also illustrated how the birth of a child with chronic illness or disability can have an adverse affect on family relationships and friendships, and how such relationships can be a source of stress and upset rather than support. The moderating effects of social support on the stressors associated with having a chronically ill or disabled child has been previously studied (e.g. Barakat and Linney, 1992; Oka and Ueda, 1998) and it has found to be beneficial to parents (Warfield et al., 1999). Equally, perceived support has been found to have a positive effect on health and well-being (e.g. Schwarzer and Leppin, 1989). A questionnaire was created for the purpose of this study to explore friend and family relationships, their importance to parents, and how they had changed since the birth of their child with MPS IH. Five questions on this measure pertaining to actual support received from family and friends, and to perceived support, were inspired by the Family Coping Study (Marks and Sykes, 2000). Additional questions were added to these, relating to the importance of friendships and family members, and how relationships have changed since the birth of the child affected by MPS IH. Social support was

applied here as a social-ecological resistance factor for both parent and child adjustment. This measure can be found in Appendix Miv.

The Principle Components Analysis revealed that the scale can be broken down into four factors. Factor 1 corresponds with the dimension of Support from Friends; Factor 2 with Support from Extended Family; Factor 3 with Perceived Support; and Factor 4 with Changes in Relationships. The percentage of variance explained by each factor was calculated as: Factor 1 = 27.1%, Factor 2 = 16.8%, Factor 3 = 14.2%, and Factor 4 = 10.8%. This explains 69 percent of the total variance. The Factor Analysis table can be found in Appendix Pix. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Support from Friends  $\alpha = .919$ , Support from Extended Family  $\alpha = .771$ , and Changes in Relationship  $\alpha = .573$ . Cronbach's Alpha could not be calculated for Factor 3, Perceived Support, as the data is binary (YES/NO). Thus, Factors 1 and 2 each achieve a satisfactory level of internal reliability, while Factor 3: Changes in Relationship, shows a low level of internal consistency for this sample (Nunally, 1967).

### Family Discord

This questionnaire was developed for the purpose of this study and explores family discord. It was applied as a social-ecological factor for parent adjustment. It comprises three sets of two-part questions. The first part of each item explores disagreements that may have taken place between the respondent and both immediate and extended family members regarding a number of issues relating to their child, their child's condition, and their child's care. These include the lack of support offered, criticisms of parenting, and the lack of understanding about the condition and the child's ongoing need for treatment. The second part of each item explores unexpressed feelings toward family members as a result of the same issues. It is often the case that people feel upset, unsupported, or let down by friends and family members, or feel that others do not understand their situation, without expressing these feelings to the individuals involved. Thus, as well as recording family disagreements, it was felt important to also record feelings of disappointment or resentment, whether discord has been expressed or not, as both can have a negative impact on well-being and coping.

The issues raised in this measure are valid as they reflect parents' feelings expressed during qualitative interviewing and have been illustrated in the literature as causing distress to parents and hindering coping when they have a chronically ill or disabled child (e.g. Ray, 2002; Silver, Bauman and Weiss, 1999). Each item asks parents to rate using a 4-point scale from 'none' to 'very much' how much disagreement they have had with family members, extended and immediate, about various issues relating to their child affected by MPS IH. They are then asked to rate using the same scale how much upset, disappointment, or resentment they have felt towards family members with regard to each issue, whether they have expressed their feelings or not. This line of questioning was inspired by the Family Coping Study (Marks and Sykes, 2000), in which carers of people affected by Alzheimer's Disease were asked to rate how much disagreement they had had with family members about such issues as them not spending enough time with their sick relative, not having enough patience and respect for the sick relative, and not visiting or helping out enough with the care of the sick relative. The Family Discord measure can be found in Appendix Mv.

Items were split into "disagreement items" and "disappointment items", and each set of items was subjected to factor analysis. Since bone marrow transplant is not relevant to the "MPS other" group or to the "Healthy Child" group, factor analysis was limited to the MPS IH group only.

The Principle Components Analysis for the Disagreements element of this measure revealed that the scale can be broken down into four factors. Factor 1 corresponds with Extended Family Support and Understanding of Child's Behaviour and Limitations; Factor 2 with Immediate Family Support and Understanding of Child's Behaviour and Limitations; Factor 3 with Issues relating to Child's Medical Condition and Treatment; and Factor 4 with Issues Relating to the Mother's Role or Ego (as parent, wife, home keeper, and carer). The percentage of variance explained by each factor was calculated as: Factor 1 = 22.0%, Factor 2 = 19.4%, Factor 3 = 17.4%, and Factor 4 = 12.7%. This explains 71.5 percent of the total variance. The Factor Analysis table for this part of the measure can be found in Appendix Px. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Extended Family Support and Understanding of Child's

Behaviour and Limitations  $\alpha = .872$ , Immediate Family Support and Understanding of Child's Behaviour and Limitations  $\alpha = .814$ , Issues Relating to Child's Medical Condition and Treatment  $\alpha = .755$ , and Issues Relating to Mother's Role  $\alpha = .599$ . Thus, Factors 1 to 3 each achieve a satisfactory level of internal reliability, while Factor 4: Issues Relating to Mother's Role, is low (Nunnally, 1967).

The Principle Components Analysis for the Disappointments element of this measure revealed that the scale can be broken down into four factors. Factor 1 corresponds with Family's Understanding of Child's Limitations and the Nature of the Condition (Extended and Immediate); Factor 2 with Family's Emotional Support Towards the Mother (Extended); Factor 3 with Issues relating to Mother's Role or Ego (Immediate and Extended), and Factor 4 with Issues Relating to the Child's Medical Condition and Treatment (Extended). The percentage of variance explained by each factor was calculated as: Factor 1 = 26.2%, Factor 2 = 23.8%, Factor 3 = 12.2%, and Factor 4 = 10.5%. This explains 73 percent of the total variance. The Factor Analysis table for this part of the measure can be found in Appendix Pxi. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Family's Understanding of Child's Limitations and the Nature of the Condition (Extended and Immediate)  $\alpha = .835$ , Family's Emotional Support Towards the Mother (Extended)  $\alpha = .884$ , Issues relating to Mother's Role or Ego (Immediate and Extended)  $\alpha = .616$ , and Issues Relating to the Child's Medical Condition and Treatment (Extended)  $\alpha = .560$ . Thus, Factors 1 and 2 each achieve a satisfactory level of internal reliability, while Factors 3 and 4 show borderline to low internal reliability for this sample (Nunnally, 1967).

#### **4.6.4.5. Measures Relating to Stress Processing Factors**

##### Parent Anxiety about Child Welfare and Perceptions of Offspring Risk

This measure was developed for use in this study and was employed to assess parent perceptions of child vulnerability. It was applied as a parent outcome measure and as a stress processing resistance factor for child adjustment outcomes. As illustrated by the literature reviewed and the qualitative data collected for the first phase of this study, parents of chronically ill children can experience a great deal of uncertainty in

relation to a number of areas in their lives and their children's lives (e.g. Cohen, 1995). In turn parental feelings of uncertainty can impact on both parent coping and child psychosocial outcomes (e.g. Mishel, 1984; Thompson and Gustafson, 1996). It was therefore felt important to explore this as a stress-processing resistance factor in the adjustment of parents and children affected by MPS IH. Two measures were therefore created to measure parental anxiety and perceptions of child risk. The first measure asked parents to rate from *I do not worry at all* to *I worry a great deal* on a 5-point Likert-type scale how much they worry about their child in certain situations or when carrying out a selection of activities. The activities and situations presented were all pertinent issues discussed with parents during the qualitative interviews. The second measure considers how much more at risk parents feel their children are when carrying out the same activities/situations as in the above measure, in comparison to a sibling when at the same age as their MPS child or another healthy child of the same age. The original scale used a 3-point Likert scale to rate parents' attitudes toward their children's risk of coming to harm. They were asked to state whether they felt their MPS child was less at risk, of equal risk or at more risk of coming to harm than a healthy peer or sibling when at the same age. This scale was amended following the first stage of piloting, as although parents used the whole range of scores on the scale, the total scores did not discriminate enough to fully illustrate parents' attitudes. This was changed to a 5-point Likert scale, using the same *less at risk* to *at more risk* parameters, so that the scores are more discriminatory. These measures can be found in Appendix Mii.

Four versions of the Anxiety/Risk scale were designed so that age appropriate behaviours could be included: 0-5 years, 6-11 years, 12-17 years, and 18+ years. However, on examination after the piloting stage, the 0-5 year and 6-11 year versions were combined to create an under-12s version, by the removal of one item from each version. The items removed were: "picking up and choking on a small object" from the 0-5 version; and "use of the internet" from the 6-11 version. These items were chosen for removal as they were the only items that differed between the measures. It was felt this measure would be more useful statistically if these age groups were combined. Similarly, and for the same reason, the 12-17 years and the 18+ years versions of this measure were also combined. The items removed were "use of the internet" from the 12-17 year version, and "living independently" from the 18+

version. For Factor Analysis purposes, only the BMT IH and Healthy Child groups' scores were used, as they all completed the revised version of the Risk Attitudes Scale with the 5-point likert scale.

The Principle Components Analysis for the under-12s version of the Parent Anxiety scale revealed that the scale can be broken down into three factors. Factor 1 corresponds with the dimension of Everyday Childhood Activities; Factor 2 with the Social Integration and Emotional Wellbeing of the Child; and Factor 3 with Child Mortality and Major Vulnerability. The percentage of variance explained by each factor was calculated as: Factor 1 = 31.8%, Factor 2 = 18.4%, and Factor 3 = 16.6%. This explains 67 percent of the total variance. The Factor Analysis table for this version of the measure can be found in Appendix Piii. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Everyday Childhood Activities  $\alpha = .832$ , Social Integration and Emotional Well Being of Child  $\alpha = .878$ , and Child Mortality and Major Vulnerability  $\alpha = .677$ . Thus indicating that Factors 1 and 2 achieve a satisfactory level of internal reliability, and that Factor 3 is borderline (Nunnally, 1967).

The Principle Components Analysis for the 12 years and over version of the Parent Anxiety scale revealed that the scale can be broken down into four factors. Factor 1 corresponds with the Worries about Adult Social Independence; Factor 2 with Worries about Physical and Emotional Vulnerability (activities parents can potentially control); Factor 3 with Loss of Child; and Factor 4 with Activities Parents Have Less Control Over. The percentage of variance explained by each factor were calculated as: Factor 1 = 28.1%, Factor 2 = 20.6%, Factor 3 = 14.2%, and Factor 4 = 14.1%. This explains 77% of the total variance. The Factor Analysis table for this version of the measure can be found in Appendix Piv. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Adult Social Independence  $\alpha = .686$ , Physical and Emotional Vulnerability (parents can control)  $\alpha = .722$ , Loss of Child  $\alpha = .434$ , and Activities Parents Have Less Control Over  $\alpha = .497$ . Thus Factors 1 and 2 achieve a borderline level of internal reliability, while Factors 3 and 4 remain low for this sample (Nunnally, 1967)

The Principle Components Analysis for the under-12s version of the Risk Perceptions Questionnaire revealed that the scale can be broken down into four factors. Factor 1 corresponds with the dimension of Risk of Physical Harm; Factor 2 with Activities Parents Have Less Control Over; Factor 3 with Risk of Emotional Harm; and Factor 4 with Loss of Child. The percentage of variance explained by each factor was calculated as: Factor 1 = 21.5%, Factor 2 = 18.2%, Factor 3 = 17.7%, and Factor 4 = 12.8%. This explains 70 percent of the total variance. The Factor Analysis table for this version of the measure can be found in Pv. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Risk of Physical Harm  $\alpha = .789$ , Activities Parents Have Less Control Over  $\alpha = .720$ , Risk of Emotional Harm  $\alpha = .760$ , and Loss of Child  $\alpha = .364$ . Thus, each of the first three subscales achieve a satisfactory level of internal reliability, while Factor 4 remains low (Nunnally, 1967).

The Principle Components Analysis of the 12 years and over version of the Risk Perceptions Questionnaire revealed that the scale can be broken down into five factors. Factor 1 corresponds with Risk of Emotional Harm; Factor 2 with Risks Associated Adult Social Independence; Factor 3 with Risk of Physical Harm; Factor 4 with Risks Associated with Physical Aspects of the Disease (physical appearance, physical functioning, and nature of the condition), and Factor 5 with Activities Parents Have Less Control Over. The percentage of variance explained by each factor were calculated as: Factor 1 = 20.1%, Factor 2 = 19.8%, Factor 3 = 16.7%, Factor 4 = 16.5%, and Factor 5 = 13.5%. This explains 87 percent of the total variance. The Factors Analysis table for this version of the measure can be found in Appendix Pvi. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Risk of Emotional Harm  $\alpha = .854$ , Risks Associated with Adult Social Independence  $\alpha = .505$ ; Risk of Physical Harm  $\alpha = -.044$ , Risks Associated with Physical Aspects of the Disease  $\alpha = .589$ , and Activities Parents Have Less Control Over  $\alpha = .853$ . Thus, Factors 1 and 5 achieve a satisfactory level of internal reliability (Nunnally, 1967). Factor 2, Risks Associated Adult Social Independence, has low internal reliability. However, with the removal of one item, "natural death", internal reliability is raised to .836. Since this item was negatively correlated with the other items in this factor, and since it seems to be incongruous with the other items, it was deleted. Furthermore, with the removal of one item,



“exclusion” from Factor 3: Risk of Physical Harm, internal reliability is raised to .873. Again, since this item seems incongruous with the other items in the factor, and is negatively correlated with them, it was removed.

### Parent Expectations of Child

As a further measure of parental perceptions of child vulnerability, again to be applied as a parent outcome measure and as a stress processing resistance factor for child adjustment, parents were asked about their future expectations of their child in a number of areas. This was a new measure developed for use in this study, which again relates to uncertainty and how it can impact on parent coping and in turn on child psychosocial outcomes. As revealed by the qualitative data many parents did not know for how long their children would live, and this was explicitly highlighted by one parent as a factor that had affected the parenting of her child. Similarly some parents stated that they felt it was unlikely their children would live independently or be able to take care of themselves in the future. This has implications for the way in which parents and children perceive and appraise the illness and the situation they are in, impacting on their coping abilities. A measure was therefore created which explores parents’ long-term expectations of their children affected by MPS I post-BMT. The measure presents a selection of activities or outcomes, and asks parents to state how likely they think it is that their child will achieve the outcomes in adulthood. Parents were asked to make their rating using a 5-point Likert-type scale from *Extremely Unlikely* to *Very Likely*. Two versions of this measure were designed: under-18s and 18 years and over. This measure can be found in Appendix Miii.

The Principle Components Analysis for the under-18s version of this measure revealed that the scale can be broken down into three factors. Factor 1 corresponds with the dimension of Hopeful Expectations of Independent Living; Factor 2 with Expectations of Interpersonal Relationships; and Factor 3 with Physical Implications of the Disease. The percentage of variance explained by each factor was calculated as: Factor 1 = 34.3%, Factor 2 = 19.9%, and Factor 3 = 15.6%. This explains 70 percent of the total variance. The Factor Analysis table for this version of the measure can be found in Appendix Pvii. Computation of the internal consistency for each of the factors using Cronbach’s Alpha revealed the following scores: Hopeful Expectations

of Independent Living  $\alpha = .884$ , Expectations of Interpersonal Relationships  $\alpha = .819$ , and Physical Implications of the Disease  $\alpha = .580$ . Thus, Factors 1 and 2 achieve a satisfactory level of internal reliability, while Factor 3 is low for this sample (Nunnally, 1967).

The Principle Components Analysis for the 18 years and over version of this measure revealed that the scale can be broken down into four factors. Factor 1 corresponds with the dimension of Expectations of Perceived Realistic Possibilities; Factor 2 with Perceived Unrealistic Expectations; Factor 3 with Unknown Quantity (Expectations Associated with Physical Aspects of the Disease); and Factor 4 with Realistic Expectations of Supported Living. The percentage of variance explained by each factor were calculated as: Factor 1 = 28.1%, Factor 2 = 27.6%, Factor 3 = 20.2%, and Factor 4 = 14.5%. This explains 90% of the total variance. The Factor Analysis table for this version of the measure can be found in Appendix Pviii. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Perceived Realistic Possibilities  $\alpha = .867$ , Perceived Unrealistic Expectations  $\alpha = .943$ , Unknown Quantity  $\alpha = -.243$ , and Realistic Expectations of Supported Living  $\alpha = .798$ . Thus, Factors 1, 2, and 4 achieve a satisfactory level of internal reliability (Nunnally, 1967). With the removal of one item, "old age" from Factor 3: Unknown Quantity, internal reliability for this Factor is raised to .886, a satisfactory level of internal reliability. This item was therefore deleted, which transforms the Factor to the dimension of Interpersonal Relationships.

### Beliefs and Attitudes

This measure was developed for the purpose of this study and applied here as a stress processing resistance factor for both parent and child adjustment. It measures religious and fatalistic beliefs and was administered to mothers as part of the survey. Positive cognitive appraisal has been purported as a vital resistance factor in parents' adaptation to having a chronically ill child (Wallander et al., 1989) and the way in which families make sense of the illness the child is affected by is fundamental to adaptation. Previous studies of families affected by chronic illness and disability have found those with the tendency to define negative life events as meaningful experiences to be better able to adjust than families unable to ascribe positive

meaning to experiences (McCubbin and McCubbin, 1993). Furthermore, people who are able to put things in a religious context have been found to cope better with negative life events than those who cannot (Venters, 1981). Indeed, religion has been found to be a key resource for some parents, and is linked to coping and internal locus of control, which allows parents to view the experience of having a chronically ill child as enhancing their own spiritual growth (Hill, 1994; Williams, 1993; Kelleher and Islam, 1996). The data from the qualitative phase of this study support these findings, as over half of the families interviewed ascribed positive meaning to their child's condition and their existence in the world. Religious and fatalistic beliefs were also expounded by three of the eight families interviewed, which they linked to their coping efforts and their child's survival. A measure was therefore developed for use in this study which explores parents' religious or fatalistic beliefs, which was administered to mothers. It comprises four items which ask parents to rate the strength of their religious and fatalistic beliefs on 5-point scales from none or never to very much or very often. The first question asks specifically about the strength of the respondent's religious beliefs, the second asks to which degree the respondent believes in fate, the third asks how often the respondent has felt that someone or something spiritual were looking out for them, and the fourth asks how much the respondent feels luck has played a role in getting them where they are today. These questions can be found in the Survey Schedule in Appendix I).

The Principle Components Analysis revealed that the scale has one factor with eigen values higher than 1.0. The solution could therefore not be rotated, and the component matrix is shown in the Factor Analysis table presented in Appendix Pxii. Factor 1 corresponds with the dimension of Religious or Fatalistic Beliefs. This factor explains 51 percent of the variance. Computation of the internal consistency for this measure using Cronbach's Alpha revealed the following scores: Beliefs and Attitudes  $\alpha = .674$ . However, with the removal of item 4, which pertains to "luck", Cronbach's Alpha is raised to .747. Since Luck in this case does not have the same spiritual connotations that the other three items have, and it has a factor loading below 0.4, it was removed from the scale. Thus, without this item the Beliefs and Attitudes Scale achieves a satisfactory level of internal reliability (Nunnally, 1967).

### Satisfaction with Schooling

Since children from the age of four years spend a significant amount of their lives in the school environment, it was felt important that the school experience of this sample of children be explored. This information was gathered to reflect a finding of the qualitative phase of this study and of the literature reviewed, which illustrated how the lack of understanding of the child's needs in the school setting can cause distress for parents and can have implications for both parent and child adjustment. Issues associated with schooling are seen as pertinent to the social and emotional development of this group of children and young people. This not only involves peer relationships, it also has implications for teacher-child interaction, and the level and appropriateness of learning support for those who require it. Thus, parents were asked to rate from 1-7, "not at all satisfied" to "extremely satisfied", how happy they have been with their child's schooling from an academic perspective, a social perspective, and an emotional perspective. This measure can be found in Appendix Mvii.

The Principle Components Analysis revealed that the scale has only one factor with eigenvalues higher than 1.0. The solution could therefore not be rotated, and the component matrix is shown in the Factor Analysis table which is presented in Appendix Pxiii. Factor 1 corresponds with the dimension of Parental Satisfaction with Child's Schooling. This factor explains 80 percent of the variance. Computation of the internal consistency for this measure using Cronbach's Alpha revealed the following scores: Parental Satisfaction with Child's Schooling  $\alpha = .847$ . Thus, this measure achieves a satisfactory level of internal reliability (Nunnally, 1967)

### BMT Perceptions

Drawing from Green and Solnit's 'A Vulnerable Child Syndrome' (1964) as discussed in the literature review, a set of questions was created for the purpose of this study to record mothers' perceptions of the bone marrow transplant experience. The aim was to gauge how practically problematic, emotionally challenging, and medically difficult the process was perceived to be, and in particular how the mother perceived her baby's experience to be from both an emotional and medical perspective. This line of questioning was designed to act as a further measure of

maternal perception of child vulnerability, and was applied here as a stress processing variable. As illustrated by the literature reviewed perceptions of child vulnerability to illness or death have been shown to have implications for parents' cognitive appraisal of their coping abilities (e.g. Kazak et al., 2004). It has also been linked to a number of outcomes for children that could interfere with their adjustment (e.g. Anthony et al., 2003; Bendell et al., 1994). A set of four questions was asked, which required the mother to rate how challenging the BMT experience was, using pre-coded responses, which were developed following qualitative interviewing. These can be found in the Survey Schedule in Appendix I.

The Principle Components Analysis revealed that the scale can be broken down into two factors. Factor 1 corresponds with Aspects of the BMT process that are tangible, i.e. medical and practical; and Factor 2 corresponds with Emotional Involvement for both the mother and child. The percentage of variance explained by each factor was calculated as: Factor 1 = 36.4%, and Factor 2 = 30.1%. This explained 66 percent of the total variance. The Factor Analysis table can be found in Appendix Pxiv. Computation of the internal consistency for each of the factors using Cronbach's Alpha revealed the following scores: Tangible Factors  $\alpha = .522$ ; and Emotional Involvement  $\alpha = .390$ . Thus, both Factors achieve low internal reliability for this sample.

#### **4.6.5. Measures Completed by Fathers**

Parenting Stress Index (Abidin, 1995)

See point 4.6.4.3 on page 204 for details.

Sense of Coherence (SOC) (Antonovsky, 1979)

See point 4.6.4.3 on page 205 for details.

MPS I Health Assessment Questionnaire – Part Two

See point 4.6.2. on pages 190-191 for details.

#### **4.6.6. Data Computation**

1. Three discrepancy variables were created: between mother's and father's scores of Sense of Coherence, Parenting Stress Index, and Level of Assistance Required by Patient. In order to do so, fathers' scores were subtracted from mothers' scores, which showed the difference between the scores. This was carried out on overall scores only. The purpose of this process was to create variables which illustrated the degree to which mothers and fathers had a shared or discrepant view of parenting stress and coping, and of their child's care needs. These were applied as a social-ecological resistance factors, and reflect the findings of the qualitative phase of this study and the literature reviewed, which illustrate how issues of support and understanding between spouses can impact upon parent adjustment, family functioning, and in turn on child psychosocial outcomes.

2. Three variables were created to represent some of the medical information gathered from parents:

a) MED1 represented the number of visits or telephone calls to the GP, Consultant, and Casualty as reported by parents as follows: Number of telephone phone calls made to the GP in the last 6 months + Number of visits to the GP in the last 6 months + Number of telephone calls to the local Consultant (Paediatrician) in the last 6 months + Number of telephone calls to the MPS consultant in the last 6 months + Number of visits to casualty in the child's lifetime. The rationale for collecting this information is given in point 4.6.2. Disease and Disability Factors, under the heading patient medical history. The data were summed by adding the total number of medical visits reported together for each participant.

b) MED 2 represents the number of visits made to hospitals as follows: Number of visits to the local hospital for routine checks in the last 12 months + Number of visits to the MPS specialist hospital for routine checks in the last 12 months. The data were summed by adding the total number of visits made together for each participant.

c) MED 3 represents the number of nights spent in hospital as follows: Number of nights spent in hospital in lifetime + Number of nights spent in hospital during 1<sup>st</sup> BMT + Number of nights spent in hospital during 2<sup>nd</sup> BMT + Number of nights spent in ICU. The data were summed by adding the total the total number of nights spent in hospital for each participant.

3. An objective measure of BMT complications was created as follows: Severity of BMT complications + Severity of GvHD + Ongoing complications related to GvHD and their severity.
4. A measure of family utilitarian resources (SES2) was created as follows: Mother's education + father's education + mother's NSSEC + father's NSSEC + gross annual income. Weighted means were used to account for single parent families. NSSEC scores were reversed to match the direction of education and income scales.
5. Patient Cognitive Function was calculated in terms of standard deviations from the mean, as the normative means and standard deviations differed between the instruments used (Griffiths, WISC-III and WAIS-III). Each instrument has a standardised mean and respective standard deviation, thus total measure scores were recalculated to reflect the number of standard deviations they differed from their respective normative mean. Examination of subscale scores for each of these measures can be found in Chapter Seven.

## CHAPTER FIVE

### RESULTS

### DEMOGRAPHICS

#### **5.0. Demographics**

This chapter examines parent participant demographics in relation to age, ethnicity, education level, work situation, job classification, and income. It also examines patient participant demographics in relation to the bone marrow transplant, medical history, and MPS genotype. Information presented includes the number of transplants the individual had, their age at transplant, their relationship to the bone marrow donor, the type of transplant undergone, whether or not the individual experienced complications during the transplant and continues to do so, and mothers' perceptions of the bone marrow transplant experience. Though not demographic, mothers' perceptions of the bone marrow transplant experience are included here as they did not fit with the study's model as an outcome, but give context to mothers' perceptions of the difficulties patient participants experienced during and after the transplant.

#### **5.1 Family Demographics and Patient Medical Information**

Demographic information was collected for all parent participants, including mothers and fathers who lived together as part of the family, either married or cohabiting. It includes information collected about the three fathers who chose not to complete questionnaires, and excludes the biological father who completed questionnaires but no longer lives as part of the family. Thus the following demographic information describes 44 mothers and 38 fathers. All parents were aged between 21 and 70 years. Predominantly they were white Caucasians and their first language English. A Punjabi interpreter was required to interview one mother. See tables 5-1 to 5-3 below.



**Table 5-1 Age of Parent Participants**

Years of Age	Mother	Father
21-30 years	9	4
31-40 years	20	17
41-50 years	8	8
51-60 years	6	9
61-70 years	1	0
TOTAL	44	38

**Table 5-2. Ethnicity of Parent Participants**

Ethnicity	Mother	Father
White	36	32
Irish	4	4
White Other	1	1
Black	1	0
Caribbean		
Bangladeshi	2	1
TOTAL	44	38

**Table 5-3 First Language Spoken by Parent Participants**

First Language	Mother	Father
English	41	37
German	1	0
Punjabi	2	1
TOTAL	44	38

Thirty-four percent of mothers were educated above secondary school level. The remainder either left school without qualifications or were educated to secondary school level. Eighteen percent were educated to degree level or above. Fifty-three percent of fathers either left school with no qualifications or were educated to secondary school level. Forty-five percent were educated above secondary school level, with 13% having been educated to degree level or above. See table 5-4 below.

**Table 5-4 Parents' Highest Level of Education Attained**

Highest Level of Education	Mother	Father
No Qualifications	10	7
Secondary School Qualification	19	13
Higher Education	1	1
Further Education and HND (Vocational)	6	11
Degree of Equivalent other	4	3
Postgraduate, Master, Diploma, Doctorate	4	2
Not known	0	1
<b>TOTAL</b>	<b>44</b>	<b>38</b>

Thirty-four percent of mothers worked either full- or part-time. Seventy-four percent of fathers worked either full- or part-time. Mothers that did not work and classed themselves as 'at home looking after the family' were classified as 'supported' if they were financially supported by a current or ex-partner. Mothers who did not work and who were either single or had partners who were out of work were classified as 'at home looking after the family – unsupported'. See table 5-5 below for details of mothers and fathers' employment status.

**Table 5-5 Parents' Current Work Situation**

Current Work Situation	Mother	Father
Full-time	3	27
Part-time	12	1
At home looking after family/ Supported	16	0
At home looking after family/ Unsupported	11	0
Retired	1	1
Long-term sick leave	1	0
Available for work/Job seeking	0	8
Other	0	1
<b>TOTAL</b>	<b>44</b>	<b>38</b>

Mothers and fathers current, or previous job if not currently in paid employment, were classified using the National Statistics Socio-Economic Classification 2004. See table 5-6 below for details.

**Table 5-6 NS SEC Job Classification of Mother and Father Current and/or Previous Job if Currently not in Work**

NS SEC Job Classification	Mother	Father
1 Higher Managerial and Professional Occupations	0	0
1.1 Employers and Managers in Larger Organisations	1	3
1.2 Higher Professionals	1	3
2. Lower Managerial and Professional Occupations	11	7
3. Intermediate Occupations	5	6
4. Small Employers and Own Account Workers	3	7
5. Lower Supervisory, Craft and Related Occupations	1	2
6. Semi-Routine Occupations	11	1
7. Routine Occupations	6	2
8. Long-term Unemployed/Never been in Paid Work	5	7
<b>TOTAL</b>	<b>44</b>	<b>38</b>

Twenty-six (59%) mothers did not change their work as a direct result of their child's needs. Those that did change their work situation either went part-time, gave up their jobs, or did not return to work as they had planned after maternity leave. Those that did not change their work did not necessarily work before they had their child. Thirty-two (73%) fathers did not change their work situation due to their child's needs. Thirty-three (75%) mothers took most responsibility for the day-to-day care of their child affected by MPS I. Eleven (25%) mothers equally shared the care of their child with their partner. Twenty-nine and a half percent of this study sample's gross annual income was at or below £20,000. Forty-three percent earned between £20,000 and £50,000 per annum, and 13.6% earned over £50,000. One participant did not wish to impart information pertaining to income for this research project. See Table 5-7 below for details.

**Table 5-7 Family Gross Annual Income**

Gross Annual Income (£)	Frequency
< 10,000	2
10,001-15,000	3
15,001-20,000	13
20,001-25,000	3
25,001-30,000	2
30,001-35,000	4
35,001-40,000	4
40,001-45,000	2
45,001-50,000	4
50,001-55,000	1
55,001-60,000	0
60,001-65,000	1
65,001-70,000	0
70,000 +	4
Missing	1
<b>TOTAL</b>	<b>44</b>

### 5.1.1 Historical Bone Marrow Transplant Information

Twenty-seven (61%) of the patient participants had one transplant; while the remainder went on to have two (9 female, 8 male). The age of the infants at first transplant ranged from 5 to 36 months. However, 36 months is an exception in this patient group, and this individual is considered to have a milder genotype. With the exception of this child the oldest child at first transplant was 26 months. Again, with the exception of one child, the age range of the infants at second transplant was 11 to 36 months. Thus the mean age at BMT 1 was 14.74 months (SD = 5.30), and at BMT 2 was 23 months (SD = 6.44).

Donor information for the BMT that engrafted is as follows: Eighteen (41%) were siblings, 6 (14%) were a parent, 1 (2%) was another relative, and 19 (43%) were unrelated. Six patients received enzyme replacement therapy immediately prior to and post BMT. These were all recently transplanted infants.

Six (13.5%) patients had stem cell transplants, 2 (4.5%) had cord blood transplants, and 36 (82%) had bone marrow transplants. This information was gleaned from the parents and was not medically verified.

Three mothers had two MPS pregnancies in total. Two of them terminated the second pregnancy, and one went on to have the child. One mother therefore has two children affected by the disorder. However, only one of these children participated in the research. In total, 15 (34%) of the mothers had had genetic counselling for subsequent pregnancies.

During the transplant, 20 (45%) children experienced complications. Two were considered by the parents to be 'minor', two to be 'moderate', and 16 to be 'serious'. Twenty-eight (64%) children experienced Graft versus Host Disease (GvHD). For 14 of them this was considered to be 'minor', for 4 it was considered to be 'moderate', and for 10 it was considered to be 'serious'. At the time of interview two children had ongoing problems associated with GvHD. One was oxygen dependent and had lung complications, and the other had a muscle wasting condition and had lost the ability to walk. Both were considered to be serious problems. This information was gleaned from the parents and not medically verified.

Mothers were asked to describe the bone marrow transplant experience from four different perspectives: from a medical perspective, from the child's emotional perspective, from a practical perspective, and from the mother's emotional perspective. They were asked to do so using pre-categorised response options as set out below.

The following four tables (5-8 to 5-11) illustrate the differential ways in which the families in this sample experienced their child's bone marrow transplant. As can be seen, the first question, which pertained to the child's experience from a medical perspective, prompted the most negative response, with 41% of mothers perceiving the experience as worrying problematic or life-threateningly serious. With regard to the child's emotional experience, however, only 13.6% of mothers felt their child was upset most of the time or extremely distressed. The remainder of the infants were reported as coping reasonably well. In terms of the practicalities of one's child

undergoing a bone marrow transplant, 50% of mothers perceived the experience as problematic or very problematic, with the remaining 50% seeing the situation as posing some hassles, but found it mostly manageable. Finally, with regard to the mothers' emotional experience of the BMT, only 9% of mothers felt the experience to be very distressing and difficult to cope with or emotionally devastating. The remainder either did not allow their emotions to get involved and managed the experience 'on automatic pilot' or found the experience distressing but managed and coped 'for the sake of the child'.

**Table 5-8 Mother's Perception of the BMT process: The Medical Perspective**

<b>BMT from a Medical Perspective</b>	<b>N</b>
Completely without incident, no problems at all	5
He/she was quite sick, but nothing out of the ordinary	13
Moderately OK, there were one or two minor problems	8
Quite difficult, there were worrying problems	7
Very difficult, it was touch and go whether he/she would survive	11

**Table 5-9 Mother's Perception of the BMT process: The Child's Emotional Perspective**

<b>BMT from an Emotional Perspective (Child's Emotions)</b>	<b>N</b>
No problems, he/she sailed through it	8
Not too bad, he/she didn't appear to be too disturbed	13
Child would get very upset but would cheer up quite easily	17
It wasn't very nice he/she was upset most of the time	4
It was horrendous, my child was extremely distressed	2

**Table 5-10 Mother's Perception of the BMT process: The Practical Perspective**

<b>BMT from a Practical Perspective</b>	<b>N</b>
Very Problematic	5
Problematic	17
Some Hassles	5
Manageable	13
No Problem at all	4

**Table 5-11 Mother's Perception of the BMT process: The Mother's Emotional Perspective**

<b>BMT from an Emotional Perspective (Mother's Emotions)</b>	<b>N</b>
I didn't allow my emotions to get involved	4
Stressful but I just got on with it	15
Very distressing but I managed it and coped	21
I found it very distressing and difficult to cope with	4
It was emotionally devastating and I barely coped	0

### **5.1.2. Patient Medical History**

Mothers and fathers were sent a medical history questionnaire by post prior to the researcher's visit and were asked a number of questions regarding their child's medical history and ongoing medical problems. The purpose of this is detailed under point 5.6.2 of the measures section in the previous chapter. Parents were asked to detail the number of visits or telephone calls they had made to their child's GP, paediatrician, and MPS consultant in the previous 6 months, how many routine checks their child had had at their local hospital and specialist MPS hospital in the last 12 months, how many nights their child had spent in hospital in their lifetime including during their BMTs, how many visits their child had made to casualty, the number of surgical procedures their child had undergone in total, and the number of orthopaedic surgical procedures their child had undergone in total. Descriptive statistics for the responses given are presented in Table 5-12 below.

**Table 5-12 Patient Participant Medical Checks and Surgical Procedures as Reported by the Parents**

	N	Mean	Std Dev	Min	Max
GP visits in the last 6 months	43	2.05	3.62	.00	20.0
GP telephone calls in last 6 months	43	0.53	1.48	.00	6.0
Consultant paediatrician telephone calls in last 6 months	43	0.65	1.31	.00	5.0
MPS Consultant telephone calls in last 6 months	43	1.53	4.05	.00	20.0
Routine checks at local hospital in last 12 months	43	7.32	17.7	.00	104
Routine checks at specialist MPS hospital in last 12 months	43	9.02	15.45	.00	75.0
Total number of nights spent in hospital in lifetime	43	169.56	114.69	50	547
Total number of nights spent in hospital during 1 <sup>st</sup> BMT	43	66.49	36.80	24	180
Total number of nights spent in hospital during 2 <sup>nd</sup> BMT	15	67.93	39.57	30	150
Total number of nights spent in ICU	41	19.46	42.36	0	200
Total visits to casualty in lifetime	42	1.98	1.95	.00	8.0
Total number of orthopaedic operations	43	2.07	3.06	.00	13.0
Total number of surgical procedures in lifetime	43	4.49	3.63	.00	15.0

With regard to ongoing medical problems 19 (44%) patient participants were reported by their parents as having medical problems. These ranged from asthma and eczema to under-active thyroid, GvHD on the lungs, hydrocephalus, and emphysema. Thirty-one (72%) were reported as taking medication on a regular basis, including Penicillin, Ventolin, and painkillers. Some parents however, did not report the type of medication their child took but the number they took, which ranged from one to six types of medication. Twenty-one (49%) patient participants were reported as having hearing difficulties with eight being reported as having to wear hearing aids. Twenty-four (59%) were reported as being visually impaired. Their difficulties were described as corneal clouding, optic nerve damage, and as having to wear glasses. One patient participant was described as registered blind. Fourteen (33%) were described as having mental health problems. The problems reported mainly related to anxiety, depression, and obsessive behaviour (n = 8). Three related to behavioural problems and ADHD. The remaining three were described as having eating difficulties, autistic



tendencies, and tantrums. Three patient participants were reported as taking medication for their mental health problems. Nineteen (44%) were reported as having experienced a serious adverse event. These were reported as cardiac arrests, heart and respiratory failure, a twisted bowel, 'broken lung', and a wound infection. Eight patient participants were reported as having more than one serious adverse event, with four having two events, two having three events, and two having four events. With regard to puberty, three females were reported as starting puberty early. This was reported as starting at the ages of seven, ten and eleven.

### **5.1.3. MPS Genetic Information**

As MPS IH is a recessive genetic disorder, two deleterious genetic mutations are required: one from the mother and one from the father. Table 5-13 below shows the patients' genotype with the maternal genetic mutation featuring first and the paternal mutation second. The most commonly found mutations of the  $\alpha$ -L-iduronidase gene in Europeans are W402X and Q70X (Beesley, Meaney, Greenland, Adams, Vellodi et al., 2001). This is reflected in this sample of patients. As well as these common mutations, rare mutations, which are unique to individual families can also be seen. This illustrates the genetic heterogeneity of the MPS I population, which makes it difficult to detect mutations and to predict disease severity. The P533R mutation has been associated with some cases of milder, late onset MPS I (Clarke and Scott, 1993), and is rarely seen in Europeans (Alif, Hess, Straczek, Sebbar et al., 1999), however severe disease can result in homozygotes (containing identical alleles of a specific gene) (Scott, Litjens, Nelson, Brooks et al., 1992). Table 5-13 below details the frequency of genetic mutation combinations seen in this group of patients.

**Table 5-13 MPS IH Genetic Mutation**

Genotype	Frequency
W402X/W402X	14
W402X/Q70X	4
Q70X/Q70X	3
W402X/R619X	1
W402X/A327P	4
W402X/S633L	1
W402X/T387R	1
W402X/C49delC	1
W402X/153delC	1
W402X/C664	1
W402X/P533R	1
W402X/Unknown	2
Q70X/A75T	1
T388R/W402X	1
P533R/P533R	1
L490P/L490P	1
C871delC/C871delC	1
474-2A→ G in Exon	1
4/A79vExon2	
Not known	3
A75T/A75T	1

#### 5.1.4. Conclusion

The information presented in this section demonstrates diversity in this population of families, particularly in terms of socioeconomic background, parent education and parent job classification. It also demonstrates similarities between the parents as the majority of mothers took responsibility for the day-to-day care of their child affected by MPS IH post-BMT. In terms of the child's bone marrow transplant, the majority had bone marrow transplants, over half experienced GvHD, and the bone marrow donors were mostly either siblings or were unrelated. A significant number of the patient participants were found to experience difficulties post-BMT with just under half experiencing medical problems and hearing difficulties, and over half being visually impaired. Just under half were also reported by their parents as experiencing one or more serious adverse events in their lifetime. The data presented also illustrates the emotionally stressful experience of the bone marrow transplant from the perspective of the mother.

## CHAPTER SIX

### RESULTS

#### PARENT ADJUSTMENT OUTCOMES

##### **6.0 Parent Adjustment Outcomes**

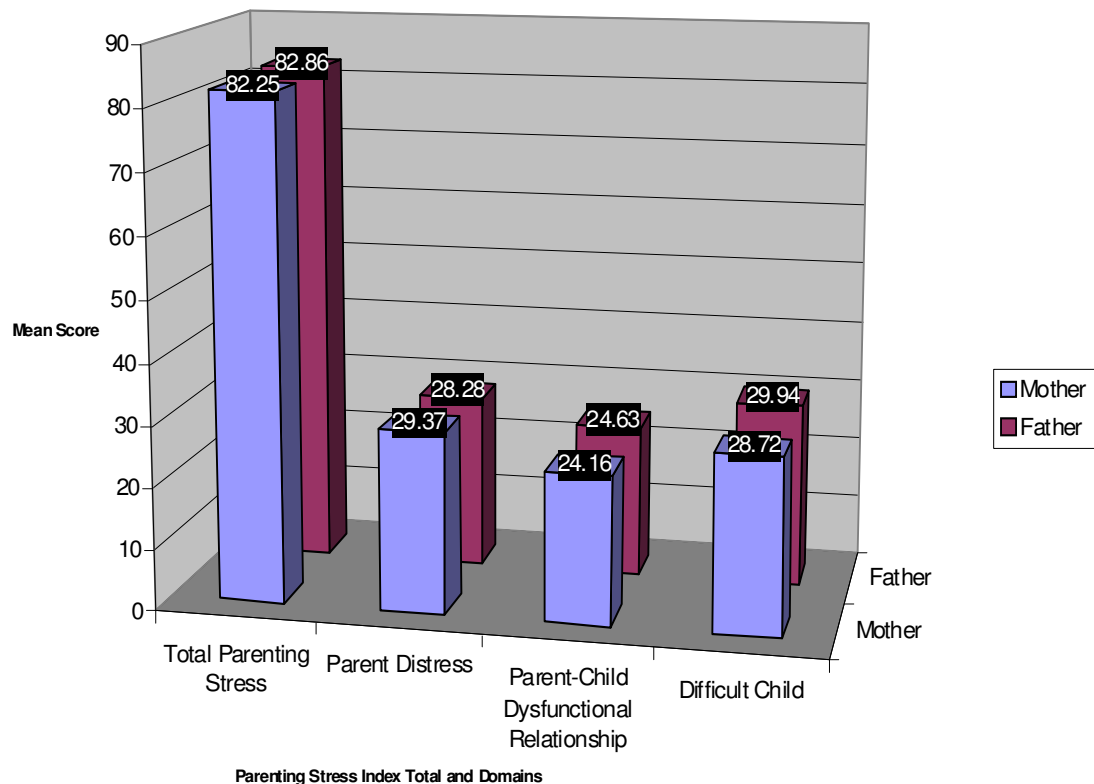
This chapter presents the findings of this study which pertain to parent participant adjustment outcomes. Findings are presented in five sections and examine parenting stress, parent coping, mothers' mental health, mothers' expectations of the child, and mothers' anxiety about child welfare and perceptions of offspring risk. Data are examined in relation to norms, between mothers and fathers, and in relation to the age of the child affected by MPS IH post-BMT. Parent outcomes are also examined using the disability stress-coping model. Thus, the moderating effect of psychosocial resistance factors on the relationship between biomedical risk factors and parent adaptation is investigated.

##### **6.1 Parenting Stress**

In order to explore the differences and/or similarities between mothers' and fathers' reports of parenting stress, independent samples t-tests were employed. Four domains of parenting stress were investigated as measured by the Parenting Stress Index (Abidin, 1995): Parent Distress, Parent-Child Dysfunctional Interaction, Difficult Child, and Total Parenting Stress. All parent participants were included in this analysis (mothers  $n = 44$ , fathers  $n = 38$ ). Figure 6-1 below shows how mothers and fathers did not differ in their reporting of parenting stress on any of the four domains. However, both mothers' and fathers' reporting of parenting stress were rather high in comparison to norms, as established by Abidin (1995). With regard to mothers' total parenting stress the scores ranged from 39 to 124, and 32.6% of mothers scored above the 90<sup>th</sup> percentile, indicating clinically significant levels of parenting stress. With

regard to fathers' total parenting stress the scores ranged from 43 to 121, and 28.6% scored above the 90<sup>th</sup> percentile, again indicating clinically significant levels of parenting stress (Abidin, 1995).

**Figure 6-1 Mother and Father Parenting Stress Index Total and Domain Mean Scores**



When breaking the sample down into child age groups (5 years and under, 6-11 years, 12 years and over) some differences between mothers' and fathers' parenting stress can be seen, although they are not statistically significant. Interestingly fathers appear to score higher than mothers for each of the age groups for Total Parenting Stress, except in the older age group, where mothers scored more highly than fathers. With regard to Parent Distress mothers and fathers of children under the age of twelve did not differ in their reports. However, the biggest difference seen between mothers and fathers Parenting Stress is for Parent Distress in the over-12s age group. Fathers' mean score of Parent Distress ( $M = 23.37$ ,  $SD = 6.21$ ) is lower than mothers' mean score ( $M = 29.45$ ,  $SD = 8.05$ ). Although the difference is not statistically significant (t

= 1.78,  $df = 17$ ,  $p = .093$ ), this could show a trend and is worthy of further investigation with a larger sample size. Similarly, with regard to Parent-Child Dysfunctional Interaction, in the under-12s age groups mothers and fathers did not differ in their reports. However, the largest difference was again found in the over-12s age group with mothers scoring higher than fathers on this domain. None of these differences were statistically significant for this domain. Finally, with regard to the Difficult Child domain of the Parenting Stress Index mothers and fathers did not differ significantly in their reporting for any of the child age groups.

Although mothers and fathers do not differ greatly from each other in their reports of parenting stress, regardless of the child's age, these findings suggest that parenting stress may differ for the individual parent depending on the age of the child and the differential stressors that are posed to parents at different developmental stages, or to different stressors associated with the family life cycle. Table 6-1 below shows how mothers differentially reported parenting stress depending on the age of their child; and how paternal stress appears to decrease for each domain on the Parenting Stress Index the older the age of the child.

**Table 6-1 Descriptive Statistics for Mothers' and Fathers' Parenting Stress Index Total and Domain Scores Across Age Group**

	Group (Mothers' N/Fathers' N)	Mothers Mean	St Dev	Fathers Mean	St Dev
Parental Distress	5 yrs and under (n = 17 + 14)	30.65	10.29	32.93	6.39
	6-11 yrs (n = 15 + 13)	27.87	8.65	26.31	6.52
	12 yrs plus (n = 11 + 8)	29.45	8.05	23.37	6.21
	Total	29.37	9.07	28.28*	7.39
Parent-Child Dysfunctional Interaction					
	5 yrs and under	24.88	4.82	26.86	5.57
	6-11 yrs	21.60	4.66	23.69	4.87
	12 yrs plus	26.54	7.22	22.25	8.60
	Total	24.16	5.70	24.63	6.26
Difficult Child					
	5 yrs and under	31.59	7.82	33.43	9.18
	6-11 yrs	26.40	6.66	28.23	6.27
	12 yrs plus	27.45	9.55	26.62	10.24
	Total	28.72	8.10	29.94	8.74
Total Parenting Stress					
	5 yrs and under	87.12	18.73	93.21	15.52
	6-11 yrs	75.87	16.36	78.23	12.19
	12 yrs plus	83.45	21.39	72.25	21.98
	Total	82.25	18.88	82.86*	17.97

\* Significant at the 0.05 level

A one-way unrelated analysis of variance showed an overall borderline significant effect for the age of the child on mother's PSI Parent-Child Dysfunctional Interaction ( $F = 2.839$ ,  $p = .070$ ). Using the Tukey HSD multiple comparison test to explore the differences between mother's parenting stress at the three levels of age, mothers reports of Parent-Child Dysfunctional Interaction were found to be greater for mothers of children aged 12 years and over than for those with children aged between 6-11 years, although this difference was not found to be statistically significant ( $p = .071$ ).

A one-way unrelated analysis of variance showed an overall significant effect for age group on father Parent Distress ( $F_{2,32} = 6.655$ ,  $p = .004$ ) and father Total Parenting Stress ( $F_{2,32} = 5.168$ ,  $p = .011$ ). Again, using the Tukey post-hoc test to explore the differences between father parenting stress at the three levels of age, fathers' reports of Parent Distress were found to be significantly greater when they had children aged 5 years and under than when they had children aged 12 years and over ( $p = .005$ ), or children aged between 6 and 11 years ( $p = .030$ ). With regard to father Total Parenting Stress the Tukey post-hoc test showed fathers of children in the lowest age group to report significantly greater stress than fathers of children aged 12 years and over ( $p = .016$ ), and than those with children aged between 6 and 11 years ( $p = .055$ ), although this difference was of borderline significance.

The above comparisons of mothers' and fathers' parenting stress illustrate how overall the parents of this patient group do not differ greatly in their reporting of stress. However, when looking at the parents in terms of the age of their children, some slight differences between mothers and fathers emerge. These differences are particularly seen amongst the parents of the children and young people in the age group 12 years and over, with mothers reporting greater Parent Distress and greater Parent-Child Dysfunctional Interaction. Although these differences are not statistically significant they suggest a possible trend that the toll exacted on the mother is greater than on the father as the child grows into adolescence and adulthood. However, mothers' stress appears to remain fairly stable across each of the three age groups, while fathers stress appears to be at its highest in the youngest age group and at its lowest in the highest age group. Thus suggesting that it is not mothers' stress that increases with the age of the child, but fathers' stress that decreases, which accounts for the difference between mothers' and fathers' reports of parenting stress in this older age group.

### **6.1.1. Predictive Association between Risk and Resistance Factors and Parenting Stress**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in parenting stress than illness and disability related risk factors. Variables were entered into the hierarchies in theoretical groups as follows: 1) illness-related risk factors, 2) psychosocial risk factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Illness-related risk factors that were added into the model for mothers' parenting stress were patient physical functioning as measured by the MPS I Health Assessment Questionnaire and the Child Health Questionnaire (Landgraf et al., 1999), cognitive function as measured by the Griffiths (Griffiths, 1971), WISC-III (Wechsler, 1992) and WAIS-III (Wechsler, 1998) developmental tests, and adaptive function as measured by the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984). Psychosocial risk factors included the number of routine medical visits to the MPS consultant in the last 12 months as measured by the Medical History Questionnaire and patient internalising problems as measured by the Behaviour Assessment System for Children Parent Report (Reynolds and Kamphaus, 1998). Stress processing variables that were added into the model were the time and emotional impact of the child's condition on the mother and mothers' perception of the child's overall health as measured by the Child Health Questionnaire. Social-ecological resistance factors that were added into the model pertained to family functioning, in particular family cohesion as measured by the Child Health Questionnaire, and mothers' and fathers' shared or discrepant view of parenting stress and the child's care needs as measured by the Parenting Stress Index and MPS Health Assessment Questionnaire Part Two, respectively. As described in point 5.6.5. of the last chapter, new variables were created to show the difference between mothers' and fathers' scores on these measures. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-2 below to show the relative contribution of each of the risk and resistance dimensions with regard to Mothers' Parenting Stress.



**Table 6-2 Model for Mothers' Total Parenting Stress**

Variable	Standardised Beta	t-value	Sig
<b>F = 6.459<sub>3,17</sub>, p = .004 (71 %)</b>			
Patient Physical Functioning	-.168	-.491	.630
Level of Assistance Required	.419	1.299	.211
Physical Functioning (2)	-.107	-.773	.450
Cognitive Function	-.129	-.843	.411
MED2: Routine Visits to MPS Hospital	-.006	-.028	.978
Mother's perception of Child Health	.045	.233	.818
Patient Adaptive Skills	-.195	-1.255	.226
Patient Internalising Problems	.108	.702	.492
Emotional Impact on Parent	.080	.496	.626
Time Impact on Parent	-.053	-.175	.863
Impact on Family	-.164	-.913	.374
Family Cohesion	-.355	-3.042	.007*
Discrepant Scores of Parenting Stress	.363	2.877	.010*
Discrepant Account of Assistance Required by the Patient	-.355	-2.507	.023*

\* Significant at the 0.05 level

With all variables entered into the analysis mothers' parenting stress was significantly predicted by the combination of the social-ecological factors pertaining to family environment, which accounted for 71% of the variance. The relationships seen indicate that mothers' parenting stress is greater when i) the family environment is less cohesive; ii) mothers' and fathers' experience of parenting stress is discrepant, i.e. when the father experiences less stress than the mother; and iii) parents' view of their child's physical needs are discrepant, i.e. when fathers perceive their child as requiring less assistance with activities of daily living than the mother perceives. Thus, family environment factors appear to moderate the effects that child illness and functional risk factors have on mothers' parenting stress.

Illness-related risk factors that were added into the model for fathers' parenting stress were patient physical functioning as measured by the MPS I Health Assessment Questionnaire and cognitive function as measured by the Griffiths, WISC-III and

WAIS-III developmental tests. Psychosocial risk factors added into the model were the level of assistance required by the child carrying out activities of daily living as measured by the MPS I Health Assessment Questionnaire, the number of routine medical visits (GP and MPS consultant) in the last 12 months as measured by the Medical History Questionnaire, and the child's global behaviour as measured by the Child Health Questionnaire. Stress processing factors that were added into the model were mother's perception of child health, time impact of the child's condition on the parent, and impact of the child's condition on the family as measured by the Child Health Questionnaire. Social-ecological resistance factors that were added into the model pertained to family functioning, in particular family cohesion as measured by the Child Health Questionnaire, mothers' and fathers' shared or discrepant view of parenting stress and the child's care needs as measured by the Parenting Stress Index and MPS Health Assessment Questionnaire Part Two, respectively, and family disagreements particularly in relation to extended family members' lack of understanding of the child and their MPS condition and support given as measured by the family discord measure designed for this study. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-3 below to show the relative contribution of each of the risk and resistance dimensions with regard to Fathers' Total Parenting Stress.

**Table 6-3 Model for Fathers' Total Parenting Stress**

Variable	Standardised Beta	t-value	Sig
<b>F = 11.075<sub>4,20</sub>, p = .000 (80%)</b>			
Patient Physical Functioning	.251	1.115	.278
Cognitive Function	-.178	-1.494	.151
Level of Assistance Required	.041	.157	.877
MED1: Calls/Visits to GP	.053	.457	.653
MED2: Routine Visits to MPS			
Hospital	-.026	-.181	.858
Mother's Perception of Child Health	-.013	-.101	.920
Child's Global Behaviour	-.163	-1.254	.224
Time Impact on Parent	.033	.238	.814
Impact on Family	-.050	-.339	.738
Family Cohesion	-.267	-2.294	.033*
Discrepant Ratings of Assistance			
Required by Patient	-.261	-1.757	.094
Discrepant Ratings of Parenting Stress	-.648	-5.391	.000**
Family Disagreements: Support and			
Understanding of Child/MPS	.224	1.863	.077
Condition			
* Significant at the 0.05 level			
** Significant at the 0.005 level			

With all variables entered into the analysis, fathers' parenting stress was significantly predicted by the combination of social-ecological factors that pertained to family functioning, accounting for 80% of the variance. However, unlike mothers, the role of extended family and their understanding and acceptance of the child also played a role for fathers. These relationships indicate that fathers' parenting stress is greater when i) there is a less cohesive family environment; ii) parents' stress is discrepant, i.e. when the father is experiencing more parenting stress than the mother; iii) parents' view of their child's physical needs are discrepant, i.e. when fathers' perceive their child to as needing more assistance than the mother perceives; and when iv) there is less support and understanding from extended family members regarding the child and the child's condition. Thus, family environment factors appear to moderate the effects that child illness and functional risk factors have on fathers' parenting stress. Some of these relationships are of borderline significance, but worthy of discussion with such a small sample size

### **6.1.2. Discussion**

The results of the present analysis illustrate how maternal parent distress is greater than that of fathers when their child is in adolescence and/or adulthood. They also show maternal parent-child dysfunctional interaction to be higher in the mothers of the older children than in the mothers of the younger children. Paternal parent distress and overall parenting stress was found to be at its highest amongst the fathers of the youngest age group of children, those aged 5 years and under. Overall the findings showed maternal parenting stress to remain the same regardless of the age of the child, but found fathers' stress to decrease as the children got older. A caveat here is that the present study is not longitudinal however, and such age effects can only be surmised. Recent longitudinal research of this nature however, has reported that although parents' concerns may change as a child grows and develops the stress of parenting a child with disabilities persists over time (Baxter et al., 1995). The findings of the present study have implications for both mothers and fathers of children of this patient group. For mothers the implications perhaps lie in her commitment to care, particularly in the context of the mother-child attachment relationship, and the burden this has on her health and psychological well-being in the long-term. This finding may also illustrate differential parenting responsibilities between mothers and fathers as the children grow into adolescents and adults. For fathers the implications perhaps lie in the initial adjustment to the role as father in the light of an MPS IH diagnosis, and to father-child bonding in the early years. This is supported by previous research, which has related fathers' Parenting Stress to role confusion and the difficulty of adapting to the parenting role (Stork, 1995), and to paternal attachment and bonding (Beckman, 1991; Cohen, 1999; Krauss, 1993).

Issues of maternal parenting stress that relate to the day-to-day care of the child and management of the condition appear to be moderated by the presence of a cohesive and supportive family environment, and to a shared view with her partner of the child's needs. This supports previous research, which relates family cohesion to parents' coping competency and cites it as being instrumental in families' acceptance of their child's disabilities (Bristol, 1984); relates family cohesion to maternal adjustment to having a child with disabilities (Warfield et al., 1999), and proposes social-ecological factors such as family functioning and spousal support to buffer the

effect that risk factors can have on parents' perceptions of stress (Wallander et al., 1989). Previous research, which highlights spousal support as being important to mothers (Ray, 2002), and predictive of maternal adjustment (McCubbin and Patterson, 1983) is also supported here.

In the case of fathers' parenting stress, as with mothers, this appears to be moderated by the presence of a cohesive family environment, and exacerbated by discrepant experiences of parenting stress between spouses. Additionally, fathers' stress is related to the acceptance and understanding of the child by extended family members. This supports previous research which has related fathers' parenting stress to the quality of the spousal relationship (Heaman, 1995), and has shown fathers to rely more on spousal support than mothers, although it has been highlighted as an important factor in family adaptation for both mothers and fathers (Bristol et al., 1988; Goldberg et al., 1986; Grant and Whittell, 2000; Saloviita et al., 2003). It also supports research that has highlighted fathers' disappointment at family members' discomfort with their child with disabilities (Pelchat et al., 2003) and family members' lack of understanding (Ray, 2002).

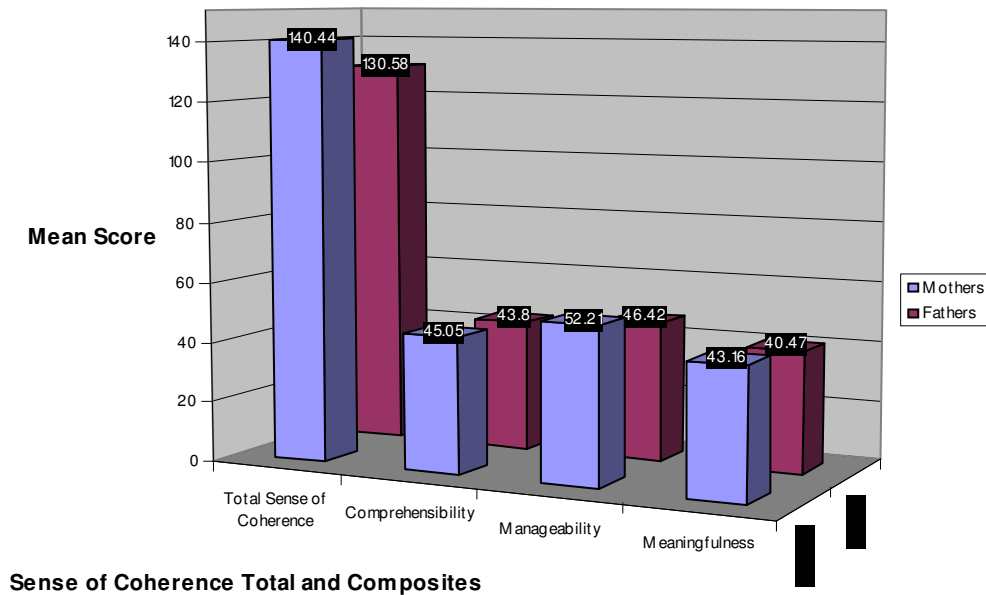
In summary then, the findings of the present study show mothers' parenting stress to not differ greatly between those with young children, middle-aged children, and adolescents/young adults. However, there is a possible trend, which suggests that those with older children experience greater parent-child dysfunction interaction than do fathers. Conversely, fathers' parenting stress is reported differently depending on the age of the child, with those with younger children reporting greater parent distress than those with older children. These findings draw attention to the early mother-child bond and fathers' early adaptation to having a chronically ill child. They also draw attention to the longer-term commitment to care given, especially by mothers. While both mothers and fathers reported relatively high Parenting stress in relation to norms, the social-ecological variables of family environment and a shared view of the child's needs between parents stood out as moderators of the effects of risk factors on perceptions of parenting stress for both parents.

These findings highlight the need for longitudinal research so that parenting stress can be explored over time. As the condition continues to be progressive in terms of physical functioning and thus the child has increasing care requirements, it would be useful to investigate the ways in which different aspects of the child's condition differentially contribute to parenting stress as the children and young people go through different developmental stages. This would enable support groups and medical professionals to better understand family dynamics and consequently provide the appropriate support. The importance of exploring the experiences of both mothers and fathers has certainly been highlighted here, and thus it is important that both parents where possible are included in future research of this nature. Attention also needs to be given to fathers' involvement with the child and adaptation to their diagnosis in the early years and to maternal involvement with the child throughout the child's life. Particular attention also needs to be given to the spousal relationship and the functioning of the family, as the moderating effects of such social-ecological factors have been demonstrated as being essential to parents' perceptions of the stresses associated with having a child affected by MPS IH post-BMT. Now that more is known about the physical outcomes of BMT for this patient group it is possible for medical professionals to give parents a clearer idea of what to expect, especially in terms of life expectancy, which may help parents to have a more long-term view of parenting. When dealing with families from diagnosis and throughout the child's life, through the educational, medical, and social challenges that are faced, it is important to work through issues with both parents, ensuring that their individual needs are met along with the child's. Support programmes that focus on the family environment would help to build a cohesive base from which parents can work.

## **6.2. Sense of Coherence**

All parent participants were included in this analysis (mothers  $n = 44$ , fathers  $n = 38$ ), which describes parents' coping abilities as measured by the Sense of Coherence Scale (Antonovsky, 1979). Of the mothers and fathers that completed this scale a number of differences emerged between the parents. Independent samples t-tests were employed to test these differences. Fathers scored lower than mothers overall on the total SOC scale, although this difference was not significant. However, the mean score for fathers ( $M = 46.42$ ,  $SD = 9.97$ ) was significantly lower ( $t = 2.48$ ,  $df = 77$ ,  $p = .015$ ) than the mean score for mothers ( $M = 52.21$ ,  $sd = 10.64$ ) on the Manageability dimension, indicating that fathers may be less able than mothers to avail themselves of, or utilise, resources to help them to cope adequately with the stressors in their lives. This is illustrated in Figure 6-2 below. In comparison to normative data, compiled by Antonovsky (1993), fathers scored quite low in comparison to male controls, but within one standard deviation of the mean score obtained by fathers of disabled children. With regard to mothers, their scores did not differ greatly from those of female controls and those of mothers of children with disabilities on the total SOC scale.

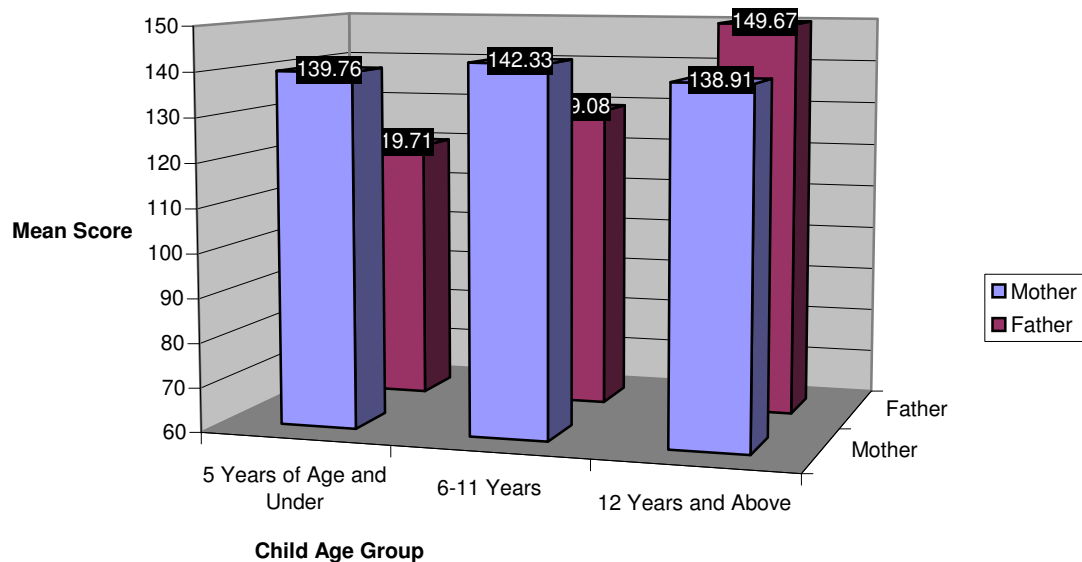
**Figure 6-2. Differences Between Mothers' and Fathers' Sense of Coherence: Total Measure Scores and Individual Dimension Scores**



When the sample is broken down into child age groups, it is clear that fathers' SOC is depleted in comparison to mothers' SOC, especially when the child is aged under-12 years. Computation of an independent samples t-test showed there to be a borderline significant difference between mothers' ( $M = 139.76$ ,  $SD = 33.18$ ) and fathers' ( $M = 119.71$ ,  $SD = 22.61$ ) mean scores of SOC when they have a child 5 years of age or under ( $t = 1.92$ ,  $df = 29$ ,  $p = .065$ ). The fathers of children in the oldest age group appear to have higher SOC than the fathers of the children in the youngest age group, while mothers' SOC remains stable regardless of the age of the child. None of these differences between mothers and fathers are statistically significant, but the stability of mothers' SOC, and the apparent rise in fathers' SOC, across age group is illustrated in Figure 6-3 below.



**Figure 6-3 Mothers' and Fathers' Mean Scores for Sense of Coherence Across Age Group**



With regard to the individual dimensions of the Sense of Coherence Scale computation of independent samples t-tests revealed some differences between mothers' and fathers' mean scores depending on the age of the child. In the case of the Comprehensibility dimension of SOC, no significant differences were found between mothers' and fathers' scores at any of the age group levels. However, again it appears that while mothers' scores remained stable regardless of the age of the child, fathers of the children in the oldest age group appear to have scored higher than fathers of the children in the youngest age group. With regard to the Manageability dimension of this scale however, fathers and mothers were found to score significantly differently when they had a child aged 5 years or under. Fathers' mean scores of SOC Manageability ( $M = 42.57$ ,  $SD = 9.38$ ) were significantly lower ( $t = 2.501$ ,  $df = 29$ ,  $p = .018$ ) than mothers' mean scores of the same domain ( $M = 52.70$ ,  $SD = 12.53$ ). There is also a borderline significant difference between mothers' and fathers' scores at the 6-11 age group level. Similarly, fathers' mean scores of SOC Manageability ( $M = 46.08$ ,  $SD = 7.65$ ) are borderline significantly lower ( $t = 1.83$ ,  $df$

= 26, two-tailed  $p = .079$ ) than mothers' mean scores of the same dimension ( $M = 52.80$ ,  $df = 11.17$ ) when they have a child aged 6-11 years. As with the total scale scores for Sense of Coherence and the Comprehensibility dimension, fathers' scores on the Manageability dimension again appear to be higher when they have children in the oldest age group compared to fathers of children in the lowest age group, while mothers' scores remain stable regardless of the age of the child. Finally, with regard to the SOC Meaningfulness dimension no significant differences were found between mothers' and fathers' scores for any of the age groups, and both mothers' and fathers' scores appear to remain stable regardless of the child's age.

As the data shows, mothers' mean scores of SOC and individual SOC dimension scores (comprehensibility, manageability, meaningfulness) do not appear to differ depending on the age of the child. A one-way unrelated analysis of variance confirmed that there is no effect for age group on mothers' Sense of Coherence on any dimension. In the case of fathers however, their mean SOC scores differ depending on the age of their child on all dimensions, including the total score, with the exception of the Meaningfulness component. A one-way unrelated analysis of variance showed an overall significant effect for age group on fathers' Total Sense of Coherence ( $F_{2,33} = 4.959$ ,  $p = .013$ ), father SOC Comprehensibility ( $F_{2,33} = 7.262$ ,  $p = .002$ ), and father SOC Manageability ( $F_{2,33} = 3.337$ ,  $p = .048$ ). The Tukey post-hoc test revealed that fathers' mean SOC scores were significantly lower when they had children aged under-5 years in comparison to those with children aged 12 years and over ( $p = .010$ ). With regard to father SOC Comprehensibility, fathers of children aged 5 years and under scored significantly lower than fathers of children aged 12 years and over ( $p = .002$ ). Furthermore, fathers of children aged between 6 and 11 years scored significantly lower than those with children aged between 12 years and over ( $p = .032$ ). Concerning SOC Manageability, again fathers of children aged 5 years and under scored significantly lower than fathers of children 12 years of age and over ( $p = .038$ ). Thus, with the exception of SOC Meaningfulness, these findings indicate that fathers' SOC in general, and fathers' Comprehensibility and Manageability are at their lowest when the child is aged 5 years and under, and at their highest when the child is over 12 years of age.

### **6.2.1. Predictive Association between Risk and Resistance Factors and Parents' Sense of Coherence**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in parent SOC than illness and disability related risk factors. Variables were entered into the hierarchies in theoretical groups as follows: 1) illness-related risk factors, 2) psychosocial risk factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Illness-related risk factors that were added into the model for mothers' SOC were patient physical functioning as measured by the MPS I Health Assessment Questionnaire and cognitive function as measured by the Griffiths, WISC-III and WAIS-III developmental tests. Psychosocial risk factors added into the model were the level of assistance required as measured by the MPS I Health Assessment Questionnaire. Stress processing factors that were added into the model were the impact the child's condition was perceived as having on the family, the emotional impact the child's condition had on the mother, and mothers' perception of the child's global health as measured by the Child Health Questionnaire. Social-ecological resistance factors that were added into the model pertained to family functioning, in particular family cohesion as measured by the Child Health Questionnaire, mothers' and fathers' shared or discrepant views of parenting stress and the child's care needs as measured by the Parenting Stress Index and MPS Health Assessment Questionnaire Part Two respectively, and family disagreements. A stress processing variable that was added into the model was mothers' parenting distress as measured by the Parenting Stress Index. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-4 below to show the relative contribution of each of the risk and resistance dimensions with regard to Mothers' Sense of Coherence.

**Table 6-4 Model for Mothers' Sense of Coherence**

Variable	Standardised Beta	t-value	Sig
<b>F = 14.282<sub>1,22</sub>, p = .001 (89%)</b>			
Patient Physical Functioning	-.308	-1.799	.086
Cognitive Function	.114	1.746	.095
Level of Assistance Required	.541	3.030	.006*
Mother's Perception of Child Health	-.152	-2.020	.056
Impact on Family	.234	2.524	.019*
Emotional Impact on Mother	-.047	-.607	.550
Family Cohesion	.033	.368	.716
Discrepant Reports of Assistance Required by Child	-.028	-.401	.692
Discrepant Reports of Parenting Stress	-.240	-2.704	.013*
Maternal Parent Distress	-.421	-3.181	.004**
Family Disagreements: Support and Understanding of Child/MPS Condition	-.339	-3.779	.001**

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

With all variables entered into the analysis, mothers' overall SOC was significantly predicted by the combination of risk, social-ecological, and stress processing factors, accounting for 89% of the variance. The relationships seen indicate that mothers' SOC is higher when the child requires more assistance with activities of daily living and when the child's overall health is perceived by the mother as poor (though this relationship was of borderline significance). Maternal SOC was also higher the less impact the child's condition was perceived as having on the family, when fathers experienced more parenting stress than mothers, when mothers reported lower parenting distress, and when there was a lower incidence of family disagreements. For the most part then, social-ecological and stress processing factors appear to moderate the effects of child illness and functional risk factors on mothers' sense of coherence. This is with the exception of the level of assistance required by the child, though this is positively related to SOC.

Illness-related risk factors that were added into the model for fathers' SOC stress were patient physical functioning as measured by the MPS I Health Assessment Questionnaire and cognitive function as measured by the Griffiths, WISC-III and WAIS-III developmental tests. Psychosocial risk factors that were added into the model were the level of assistance required by the child carrying out activities of daily living as measured by the MPS I Health Assessment Questionnaire. Stress processing factors that were added into the model were the perceived impact the child's condition had on the family and maternal perceptions of child health as measured by the Child Health Questionnaire, and mothers' parenting distress as measured by the Parenting Stress Index. Social-ecological resistance factors that were added into the model pertained to family functioning, in particular family cohesion as measured by the Child Health Questionnaire, mothers' and fathers' shared or discrepant view of parenting stress as measured by the Parenting Stress Index, and family disagreements as measured by the Family Discord measured developed for this study. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-5 below to show the relative contribution of each of the risk and resistance dimensions with regard to Fathers' Sense of Coherence.

**Table 6-5 Model for Fathers' Sense of Coherence**

Variable	Standardised Beta	t-value	Sig
<b>F = 4.827<sub>1,24</sub>, p = .038 (57%)</b>			
Patient Physical Functioning	-.182	-.814	.424
Cognitive Function	.219	1.614	.120
Level of Assistance Required	.118	.535	.598
Mother's Perception of Child Health	-.158	-.955	.349
Impact on Family	.587	2.992	.006*
Family Cohesion	-.172	-.916	.369
Discrepant Reports of Parenting Stress	.906	5.031	.000**
Maternal Mental Distress	-.378	-1.409	.172
Family Disagreements: Issues Relating to Mother's Role	-.423	-2.197	.038*

\* Significant at the 0.05 level

\* Significant at the 0.005 level

With all variables entered into the analysis, fathers' SOC was significantly predicted by the combination of social-ecological factors that pertained to family functioning, accounting for 57% of the variance. The Family Disagreements factor, which is significantly negatively related to fathers' SOC, pertains to disagreements about issues relating to the mother's role. This comprises two items, which measure the amount of disagreements that have taken place surrounding the lack of support the mother gets from immediate family members around the home, and to disagreements that stem from criticism from extended family members about the mother's parenting skills. Mothers' and fathers' discrepant parenting stress is also related to fathers' SOC, the SOC being depleted when mothers' stress is greater than fathers'. Fathers' SOC was also significantly predicted by a stress processing factor, the degree to which mothers' perceived the child's condition to impact on the family. It was found to be greater the lower the impact the child's condition was perceived to have on the family. These findings suggest that fathers' SOC is related to aspects of the home and the mother. Thus, how comprehensible, meaningful, and manageable fathers find the world appears to be related to the impact that the child's condition has on the equilibrium of the home. Individual aspects of the child's condition thus appear to be moderated by these social-ecological and stress processing factors.

### **6.2.2. Discussion**

Although parenting stress was found to be quite high for this sample of mothers in comparison to norms, their Sense of Coherence scores did not differ greatly from those of norms (Antonovsky, 1993). Moreover, Sense of Coherence scores on all dimensions of the scale were not found to differ between mothers of young children, middle-aged children, and adolescents and adults. This suggests that, while mothers experience additional stresses in their lives as a result of having a child affected by MPS IH, they have a strong Sense of Coherence, which enables them to see the world as comprehensible, manageable, and meaningful. Indeed, maternal parent distress in the present study was negatively related to mothers' SOC. This supports previous research, which has found some mothers of children with disabilities to experience high levels of stress, while maintaining a positive outlook regarding their child's adaptation and thus perceive their parental role as a positive challenge rather than distressing (Pelchat et al., 1999a; 1999b). Furthermore, mothers' SOC was positively

related to the level of assistance the child required carrying out activities of daily living and to poorer perceptions of child health, though the relationship between SOC and this latter variable was of borderline significance. These relationships suggest mothers' SOC in the present sample to be linked to her role as mother, which is nurturing and protective, meeting the child's immediate needs. This may provide the mother with a sense of meaningfulness, which helps her to view the parenting role as a worthwhile challenge (Young et al., 2002). This suggests that mothers' appraisal of the parenting role as rewarding buffers the effects of stress on adjustment.

Additionally important to mothers' SOC in the present study was the family environment and her appraisal of the impact the child's condition had on the family. Maternal SOC was found to be higher the less the child's condition was perceived as impacting on the family. This supports previous research which has highlighted mothers' appraisals of the impact the child's condition has on the family as a mediating process through which illness or functional severity may lead to parental psychological problems (e.g. Wallander et al., 1989; Ireys and Silver, 1996; Lustig et al., 1996). Social-ecological factors such as aspects of the family environment were also found to impact on maternal SOC, particularly discrepant experiences of parenting stress between mothers and fathers and the impact that disagreements had within the family. Disagreements with extended family members concerning their lack of understanding of the child's MPS condition and on-going needs, and their failure to offer support was negatively related to maternal SOC. This kind of help is important in assisting mothers to develop 'normalised' feelings about their child, and to help her make sense of the child's condition (Green, 2001). Indeed, it could be said that such support can aid mothers' SOC *comprehensibility*. Hence, if such involvement is inappropriate, insufficient, or is the cause of disagreements, mothers' ability to cope with having a child with MPS IH post-BMT may be challenged (Ray, 2002). Thus, such social-ecological and stress processing factors have been demonstrated here as moderating the effects of child illness and functional risk factors on maternal SOC.

With respect to fathers' SOC, the findings highlight their scores as being lower than mothers', particularly amongst the parents of the younger children, and particularly in the SOC Manageability domain. This is perhaps linked to the uncertain nature of the

child's future and to fathers' adjustment to his role and the child's condition in the early years. It is also possible that fathers' SOC increases as the child grows older. As time goes by there is more opportunity for the father to develop a relationship with their child and to gain a better understanding of how the condition is going to affect the child (physically and cognitively). This enables the father-child bond to develop and the father to begin to make sense of the condition and its implications for the child. Lillie (1993) observed that mothers of children with disabilities tend to be very actively involved with the child, and often excluding the father from the caring role and minimising his involvement. Since fathers' SOC in the present study is at its lowest in the group which had children aged under-5 years, and since mother SOC appears related to aspects of her role as mother, it is possible that fathers' SOC is related to the mother-child relationship and to their own role confusion, especially when the child is young. Furthermore, as the child grows older and aspects of the condition become clearer, so do ways of managing the condition. Parents become involved with decisions about their child's educational needs, and orthopaedic surgery is necessary for many to prolong mobility, improve quality of life, and minimise pain. Thus fathers may feel the situation more manageable as the child gets older and they are more involved in decisions

Similarly, contributors to fathers' SOC appear to be related to maternal appraisal, the family, and to the equilibrium of the home. In particular, fathers' SOC is higher the less the mother perceives the family to be impacted upon by the child's disabilities, and when the mother feels supported and reinforced in her role by both immediate and extended family members. While other research has related fathers' difficulties in adapting to the situation of having a child with disabilities to aspects of the disability itself (Krauss, 1993), the present study does not implicate the MPS condition per se, but rather the impact that the condition has on the family, particularly the mother. Since mothers' well-being has been related to her satisfaction in her role as parent (Beckman, 1991; Pelchat et al., 2003), it is possible that maternal satisfaction and feelings of well-being elevate paternal SOC via the spousal relationship. Indeed, as previously pointed out, research has related fathers' parenting stress to the quality of the spousal relationship (Heaman, 1995), and has shown spousal support to be important to both fathers' and mothers' adaptation (Bristol et al., 1988; Goldberg et



al., 1986; Grant and Whittell, 2000; Saloviita et al, 2003). Fathers' adjustment has also been found to be affected by the impact the child's condition has on the family (Pelchat et al., 2003) and by its time impact on the marital relationship (Heaman, 1995), and a strong predictor of distress amongst fathers is purported to be the lack of personal growth orientation within the family environment (Margalit, Leyser, Avraham, Lewy-Osin et al., 1989). Thus, social-ecological and stress processing factors appear to moderate the effects of child illness and functional risk factors on paternal SOC.

In summary then, the findings of the present study show mothers' SOC to not differ greatly between those with younger and older children. Conversely fathers' SOC was reported differently depending on the age of the child, with those with younger children reporting lower SOC than those with older children. The SOC scores thus highlight differences between mothers and fathers of this group patient group. Fathers' SOC was generally lower than mothers', particularly when the child was young and particularly in the Manageability dimension. However, while mothers' SOC did not differ depending on the age of the child, fathers' SOC was higher when they had older children, higher than mothers' SOC in the older age group. These findings draw attention to the early mother-child bond and to paternal involvement at this stage. They also draw attention to the longer-term commitment to care given, especially by mothers. The findings provide further insight into these differences by highlighting the factors that contribute to mothers and fathers' SOC. For mothers it was their role as mother, the support and understanding of the family, and their appraisal of the child's illness and its impact on the family. For fathers it was the equilibrium of the family, mother's appraisal of the child's illness and its impact on the family, and the mother's feeling that she (and the child) was supported and understood by the family, both immediate and extended. Thus, both social-ecological and stress processing resistance factors are demonstrated here as moderating the effects of child health and illness risk factors on parent Sense of Coherence. As previously mentioned, the findings of this study highlight the need for longitudinal research so that parenting stress and coping can be examined over time and individual family members' needs met from the child's diagnosis throughout the child's life.

### 6.3. Mothers' Mental Health

Computation of a one-way multivariate analysis of variance (MANOVA) with a Bonferroni adjustment applied (thus the required alpha level for 95% confidence was  $p = .017$ ), revealed no significant differences between mothers of children aged 5 years and under, 6-11 years, or 12 years and over in terms of their total and subscale mean scores on the General Health Questionnaire (GHQ) (Goldberg and Williams, 1988). A one-way repeated measures ANOVA was conducted to compare the mean subscale scores on the GHQ. The means and standard deviations are presented in Table 6-6. There was a significant effect for subscale (Wilk's Lambda = .22,  $F(5,40) = 46.93$ ,  $p = .000$ , multivariate eta squared = .78), indicating that mothers scored significantly lower on the severe depression subscale than on the somatic complaints, anxiety and insomnia, and social dysfunction subscales.

**Table 6-6 Descriptive Statistics for GHQ Total and Sub-Scale Scores Across Age Group**

	Child Age Group	N	Mean	Std Dev	Min	Max
Total Score	5 yrs and under	17	24.47	18.10	4.00	65.0
	6-11 years	15	18.87	9.48	7.00	42.0
	12 yrs and over	11	24.54	12.99	13.00	56.0
	Total	43	22.53	14.23	4.00	65.0
Somatic Complaints	5 yrs and under	17	7.88	5.41	0.00	21.0
	6-11 years	15	4.93	4.01	0.00	14.0
	12 yrs and over	11	8.0	4.88	2.00	19.0
	Total	43	6.88	4.92	0.00	21.0
Anxiety and Insomnia	5 yrs and under	17	6.70	6.23	0.00	18.0
	6-11 years	15	5.20	3.43	1.00	13.0
	12 yrs and over	11	6.0	3.46	1.00	13.0
	Total	43	6.0	4.69	0.00	18.0
Social Dysfunction	5 yrs and under	17	7.53	4.42	2.00	18.0
	6-11 years	15	7.33	1.72	4.00	12.0
	12 yrs and over	11	8.18	1.94	6.00	12.0
	Total	43	7.63	3.07	2.00	18.0
Severe Depression	5 yrs and under	17	2.47	4.54	0.00	16.0
	6-11 years	15	1.40	2.82	0.00	9.0
	12 yrs and over	11	2.36	5.10	0.00	17.0
	Total	43	2.07	4.12	0.00	17.0

### **6.3.1. Predictive Association between Risk and Resistance Factors and Mothers' Mental Health**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in mothers' mental health than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) psychosocial risk factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Illness-related risk factors that were added into the model were patient physical functioning as measured by the MPS Health Assessment Questionnaire, cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests, and adaptive functioning as measured by the Vineland Adaptive Behaviour Scales. Psychosocial risk factors added into the model were the level of assistance required by the child carrying out activities of daily living as measured by the MPS Health Assessment Questionnaire, the number of calls/visits made to the GP and MPS consultant in the last 12 months as measured by the Medical History Questionnaire, and child internalising problems as measured by the Behaviour Assessment System for Children Parent Rating Scale. The stress processing resistance factors that were added into the model were mother's perception of child's overall health as measured by the Child Health Questionnaire, maternal Sense of Coherence (SOC) as measured by the Sense of Coherence Scale (Antonovsky, 1979), maternal parenting stress as measured by the Parenting Stress Index, and mothers' perceptions of the impact the child's condition has on the family as measured by the Child Health Questionnaire. Social-ecological factors that were entered into the model were family cohesion as measured by the Child Health Questionnaire, mothers' and fathers' shared or discrepant views of parenting stress, and support from friends and extended family members as measured by the Social Support measure developed for this study. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-7 below to show the relative contribution of each of the child and family dimensions to Mothers' Mental Health.

**Table 6-7 Model for Mothers' Mental Health**

Variable	Standardised Beta	t-value	Sig
<b>F = 4.624<sub>2,17</sub>, p = .025 (71%)</b>			
Patient Physical Functioning	.010	.039	.969
Level of Assistance Required	.047	.147	.885
Mothers Perception of Child Health	-.526	-4.001	.001**
MED 1: Calls/Visits to GP	.102	.680	.506
MED 2: Calls/Visits to MPS Hospital	.480	2.810	.012*
Patient Cognitive Function	-.036	-.225	.825
Patient Internalising Problems	-.299	-1.780	.093
Patient Adaptive Skills	.082	.530	.603
Family Cohesion	-.428	-2.960	.009*
Discrepant Reports of Parenting Stress	.607	3.979	.001**
Impact on Family	.014	.078	.938
Maternal Parenting Stress	-.642	-2.964	.009*
Maternal Sense of Coherence	-.721	-2.845	.011*
Support from Friends	-.269	-2.207	.041*
Support from Extended Family	.378	2.838	.011*

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

With all variables entered into the analysis, 71% of the variance of mothers' mental health was explained and was found to be significantly predicted by the combination of an illness-related stress factor, stress processing factors relating to maternal stress and coping, and maternal perceptions of child health, and social-ecological factors relating to aspects of the family environment and spousal support; and social support. These will be discussed in relation to previous and future research, and to practical applications.

### 6.3.2. Discussion

The results show mothers' perception of overall child health and the ongoing management of the child's MPS condition to be related to mothers' mental health. In particular, mothers' perception that the child's overall health was poor and more frequent visits to the child's specialist MPS hospital were related to poorer maternal mental health with the present population. In terms of the ongoing management of the

child's MPS condition, children and young people affected by this disorder are seen at regular intervals either by their local hospital or by one of the five tertiary centres around the UK that specialise in inherited disorders of metabolism. Such visits are necessary to monitor bone and joint growth and formation, as well as height, hormones, hearing, eyesight, and neurological function. If a child or young person requires corrective surgery or has chronic on-going complications post-BMT, they are likely to have frequent visits of this nature. Thus a greater number of hospital visits in a 12 month period indicates that the child or young person is experiencing acute or chronic health problems that require management. Often families live a long distance from their specialist tertiary centre. Such visits can disrupt family life and can take the child out of school for significant amounts of time. It can be a frightening and traumatic time for the child. Moreover, if the mother takes most responsibility for the day-to-day management of the child's condition, including the organisation and management of hospital visits, more frequent visits and thus likely complications with the condition are likely to take an emotional toll on her.

Previous research has illustrated the relationship between emotional distress factors such as worry and cognitive appraisal, which can impact upon illness representations and perceived self-efficacy (Bonner et al., 2006). Such emotional distress factors have also been related to perceptions of child vulnerability, which in turn have been related to psychological distress (Stewart and Mishel, 2000). Previous research has also highlighted mothers' likelihood over fathers to take responsibility for the care of sick family members (e.g. Parks and Pilisuk, 1991) and the day-to-day management of a child's condition (e.g. Thompson et al., 1993), while simultaneously taking care of the upkeep of the home (Pelchat et al., 2003). The accumulation of such illness-related and ordinary life stressors have been highlighted as causing increased difficulties for mothers and psychosocial stress resulting from chronic illness is suggested to be a significant source of adjustment problems in mothers of chronically ill children (Manuel, 2001). The findings of the present study therefore highlight how illness-related stressors and perceptions of child vulnerability can impact on mothers' emotional adjustment, and illustrate the emotional toll that can be exacted on a mother when her child is extremely sick (Bury, 1982).

With regard to the stress processing variables of parenting stress and coping, the findings of the present study show maternal SOC to be negatively related to maternal mental health problems. This is an expected finding, which supports previous research, which has negatively correlated SOC with depression (Flannery et al., 1994; Shnyder, Buchi, Sensky and Klaghofer, 2000). The negative relationship between mothers' total parenting stress and mental health as measured by the GHQ is an anomaly however. Whilst as single variables mothers' parenting stress and total GHQ are significantly positively related in the present study, with the other child and family risk and resistance variables entered into a regression analysis, this relationship appears to reverse. It is possible that an element of parenting stress is healthy for this group of mothers, as long as it is not too discrepant with that of the father. Perhaps an element of parenting stress facilitates mothers' sense of purpose in her role. Alternatively, this result could be highlighting the child's health and general well being as the mothers' utmost priority. When the child is seriously ill or going through major surgery, in other words when he/she is perceived as extremely vulnerable by the mother, it is possible that the welfare of the child overrides any feelings of parenting stress, and that characteristics of the child and the mother's parenting experience are seen in a positive light. The job of caring for the child and ensuring the child's well being, both emotional and physical, is meaningful and purposeful to the mother and therefore not perceived as stressful. The negative relationship between maternal parenting stress and mental health could therefore be more indicative of mothers' feeling that the child is vulnerable. Her main concern is therefore the welfare of the child. Moreover, this relationship may indicate a sense of helplessness in mothers' inability to free the child from the MPS disorder and the painful and limiting complications that accompany it post-BMT. Research has found mothers of children with disabilities to become over-involved with their child (Lillie, 1993), to feel their very identities to be threatened by illness and disability in their children (Anderson & Elfert, 1989), and to become particularly emotionally involved (Pelchat et al., 2003). These issues are particularly pertinent to this parent group. Due to the nature of the condition and the plethora of uncertainties that surround every aspect of its treatment, management, and course, the perception that the child is vulnerable possibly pervades, which may have implications for mothers' emotional well-being in terms of sadness and helplessness.

Furthermore, as the findings of the present study show, mothers' mental health is also related to social-ecological factors in terms of the family environment and spousal support. The findings highlight family cohesion and mothers' and fathers' discrepant experiences of parenting stress as being strongly related to mothers' mental health. Although more mothers than fathers in the present study took most responsibility for the day-to-day care of their MPS child, they did not report greater parenting stress overall. However, it appears that when such reports are discrepant, particularly when mothers experience greater parenting stress than fathers, it has implications for mothers' mental health. This is supported by previous research which espouses the view that spousal support is more predictive of maternal adjustment to a child's disability than aspects of the child's illness or disability alone (McCubbin & Patterson, 1983; Ray, 2002), and that the lack of family support is predictive of maternal distress (Margalit et al., 1989). Research has also found mothers to be at particular risk of experiencing difficulties with parenting and adjusting to a child's disability when the family environment is not cohesive (Warfield et al., 1999). Indeed, family environments that are cohesive have been found to be more supportive (Sandler & Berrera, 1984); and parents have been found to cope better and to be able to come to terms with their child's disability more easily in a cohesive family environment (Bristol, 1984).

A further social-ecological factor relevant to mothers' mental health is social support. The findings of the present study show greater mental health problems to be related to less support from friends and more support from extended family members. It is possible that when mothers feel emotionally vulnerable they are less likely to seek help from friends, and feel more comfortable receiving assistance from parents, siblings, or the spouse's family members. Extended family members such as grandparents may also be more available than friends to provide assistance, and it may be more appropriate for a family member to provide the type of assistance that is required, which is more or less a secondary parental role. It may also be possible that when a mother is focused on a sick child, is emotionally and physically drained, and finding it difficult to cope, they are less inclined to have social contact with friends. Indeed, previous research has found grandparents to be the most regular source of help for mothers of children with illness or disability (Oka & Ueda, 1998), and such support has been found to be beneficial to mothers (Green, 2001; Pelchat et al., 2003).

However, previous research has also found experiences with extended family members to exacerbate parental stress and distress rather than alleviate it (Ray, 2002). Indeed, the receipt of support has been related to feelings of guilt and dependency (Argyle, 1993). It is possible then, with the present sample of mothers, that when child health is perceived as poor, family cohesion weak, and spousal support lacking, that assistance from extended family members exacerbates distress by creating feelings of inadequacy. If the kind of support given by extended family members does not support the mother in her role as primary carer, nurturer, and protector, it may have a detrimental affect on her mental health. In terms of friendships, it may also be possible that friends are less able to cope with illness and disability in others' children, and may feel they are unable to offer appropriate support. They may also lack the understanding necessary to deal with such intense emotions or commitment to provide support. Many of the families involved in this study reported that they found their friendships changed after the arrival of their MPS child. Indeed, it is propounded that the loss of a friendship or the support of a friend is stressful, as it is essentially the loss of a resource (Hobfoll, 1988). Poor relationships have also been related to depression, lower self-esteem and a number of physical and mental health problems (Duck, 1991).

In summary then, the ways in which parents adjust to and cope with having a child affected by MPS I is little understood. The issue is a complex one, which is worthy of attention if appropriate support is to be given and outcomes improved. These findings highlight the moderating role that stress processing factors such as appraisals and coping , and social-ecological factors such as social and spousal support, and family functioning play in the relationship between child illness and functional status factors and parent adjustment outcomes. For mothers, issues of child vulnerability are very possible here and this needs to be considered when providing support to the family, and in relation to their own as well as fathers' adjustment post-BMT. Since it is known that mothers and fathers adjust differently to having a child with a chronic health condition or disability, it is important to consider the whole family when providing support. If family functioning is instrumental in a parent's ability to cope, mothers and fathers differential needs require more attention, and need to be considered by medical professionals and service providers alike. Early intervention programmes, which encourage both parents to be actively involved in the nurturing



parental role would be beneficial to mother- and father-child attachment, and to their adjustment. Play workshops that involve the whole family would also be helpful to the development of individual family relationships with the affected child, as would the encouragement of contact with other families who have been through the same experience. This perspective advocates a shared approach to parenting together with realistic knowledge about the condition and its course post-BMT. The consequences of this in terms of parenting and subsequent child psychosocial outcomes are potentially pertinent.

#### **6.4. Parent Appraisals: Mothers' Expectations of Child Future Achievements**

All 44 mothers were included in this section of analysis which describes mothers' expectations of child future achievements and explores the variables that contribute to those expectations. Of the scenarios that mothers were presented with via the Expectations Questionnaire developed for this study, they felt their child affected by MPS IH post-BMT most likely to develop friendships, go out and socialise with friends, and go on holiday without them when they reached adulthood. They felt their child least likely to have a consummated relationship and to live completely independently with no assistance. The descriptive statistics for each item of the Expectations measure as completed by mothers of patient participants under the age of 18 are shown in Table 6-8 below.

**Table 6-8 Mothers' Expectations of Child's Future Achievements: Child Under-18 Years**

Item	N	Min	Max	Mean	Std Dev
Be in Paid Employment	36	2.00	5.00	3.80	.85
Manage Own Finances	36	1.00	5.00	3.25	1.10
Develop Friendships	36	3.00	5.00	4.47	.74
Go out and Socialise	36	2.00	5.00	4.19	.92
Have a Romantic Relationship	36	2.00	5.00	3.61	.99
Have a Consummated Relationship	36	1.00	5.00	2.80	1.19
Drive a Car or Motorbike	36	1.00	5.00	3.44	1.30
Go on Holiday (possibly with organized group)	36	1.00	5.00	4.03	1.27
Take Care of Oneself (in the short-term)	36	2.00	5.00	3.72	1.06
Live Separately from Parents (with assistance or home adaptations)	36	1.00	5.00	3.67	1.24
Live Completely Independently	36	1.00	5.00	2.64	1.22
Live to Old Age (60 years +)	36	1.00	5.00	3.03	1.30

For the majority of the behaviours or activities presented, over 60% of mothers felt it a good possibility or more likely that their child would achieve them in adulthood. This was with the exception of independent living and having a consummated relationship, which less than 30% of mothers felt to be a good possibility or more likely. It was also with the exception of the management of finances and driving a vehicle, which just over 40% of mothers felt to be a good possibility or more likely; and living to old age, which 39% of mothers felt to be a good possibility or more likely. Overall then, mothers of children aged under-18 years were quite positive about their children's future achievements, although their expectations were realistically limited. They remained to be doubtful however, about issues of adult independence. These findings also illustrate how mothers continued to have low

expectations of their child's longevity in terms of length of life, despite the bone marrow transplant.

Parents of younger infants had greater expectations of their child as would be expected. Chronological age in completed months was negatively correlated with the total score on the Expectations measure (Pearson's  $r = -.284$ ,  $p = .091$ ). Although not a significant correlation it suggests a relationship between age and parental expectations of the child. It is possible that this is due to difficulties associated with the MPS condition not being apparent until the child is older, particularly those of an orthopaedic nature. Furthermore, when children are very young, it is difficult in general for parents to have realistic expectations of their future achievements in adulthood. Where the present sample of mothers is concerned, it is possible then that those with very young children were voicing their hopes rather than expectations, resulting in the scores on this scale being higher for mothers of younger children.

With regard to mothers of patient participants aged 18 years and over, the two activities they felt their child most likely to achieve were related to social activities. In particular, they felt them most likely to develop friendships and go out and socialise with friends. The activities the mothers felt their children least likely to achieve were living independently, being able to take care of themselves in the short-term, and driving a vehicle. Descriptive statistics for each item on the Expectations scale as completed by mothers of young people aged 18 years and over are shown in Table 6-9 below.

**Table 6-9 Mothers Expectations of Child's Future Achievements: Young People Aged 18 Years and Over**

Item	N	Min	Max	Mean	Std. Dev
Be in Paid Employment	8	1.00	5.00	3.25	1.58
Manage own Finances	8	1.00	5.00	2.75	1.91
Develop Friendships	8	3.00	5.00	4.50	.75
Go Out and Socialise	8	2.00	5.00	4.37	1.06
Have a Romantic Relationship	8	1.00	5.00	3.50	1.31
Have a Consummated Relationship	8	1.00	5.00	2.62	1.30
Drive a Car or Motorbike	8	1.00	4.00	1.62	1.19
Go on Holiday (possibly with an organized group)	8	1.00	5.00	3.87	1.36
Take Care of Oneself (in the short-term)	8	1.00	5.00	2.37	1.50
Live Separately from Parents (with assistance or home adaptations)	8	1.00	5.00	3.75	1.39
Live Completely Independently	8	1.00	5.00	2.00	1.85
Live to Old Age (60 years +)	8	1.00	5.00	3.00	1.31

Of the eight individuals aged 18 years of age and over, one was in paid employment; three managed their own finances; seven had established friendships; six went out to socialise with friends; two had had a romantic relationship; one had had a consummated relationship; none drove a vehicle; six had been away on holiday without their parents; one was able to take care of him or herself in the short-term; and two lived separately from their parents, with one living independently with home adaptations, the other living in residential care. Thus, for the most part, the adults in this population were dependent on their parents and lived at home. With regard to future expectations, over 60% of mothers felt it a slight possibility or less likely that their child would be able to manage their own finances, have a consummated relationship, drive a car, take care of him or herself in the short-term, live completely independently, or live into old age. As parents were getting older themselves, the expectation that their child would live separately from them at some point was

considered a good possibility or more likely by 62.5% of mothers. This was with the view that the young person would live in assisted housing or even residential care. Since the MPS patients in this age group were in adulthood, the extent to which their condition was going to progress was apparent for mothers. They were therefore able to give accurate accounts of their expectations based on their knowledge of their child's abilities and the level of care they presently required. This sets the older age group apart from the younger age group, as mothers' 'expectations' were based more on current knowledge rather than on future assumptions or hopes.

#### **6.4.1. Predictive Association between Risk and Resistance Factors and Mothers' Expectations of Child Future Achievements**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in mothers' expectations of their child's future achievements than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) psychosocial risk factors, 3) intrapersonal resistance factors, 4) stress processing resistance factors, and 5) social ecological resistance factors. Illness-related risk factors that were added into the model were patient physical functioning as measured by the MPS I Health Assessment Questionnaire, cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests, and adaptive functioning as measured by the Vineland Adaptive Behaviour Scales. Psychosocial risk factors added were the level of assistance required by the child carrying out activities of daily living as measured by the MPS Health Assessment Questionnaire. One intrapersonal factor added into the model was maternal distress, a measure of maternal mental health as measured by the CAPI (Milner, 1986). Stress processing resistance factors that were added into the model were mothers' perception of child health, maternal SOC *comprehensibility* as measured by the Sense of Coherence Scale and maternal parenting distress as measured by the Parenting Stress Index. Social-ecological factors that were entered into the model were family cohesion and organisation as measured by the Family Environment Scale (Moos and Moos, 2002), and mothers and fathers discrepant or shared coping abilities as measured by the Sense of Coherence Scale. This analysis was restricted to mothers of children aged under-18 years of age as their children had not yet reached adulthood

and their expectations were possibly less clear cut than those of the mothers of the adults. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-10 below to show the relative contribution of each of the child and family dimensions to Mothers' Expectations of child future achievements.

**Table 6-10 Model for Mothers' Expectations of Child Future Achievements: Child Under 18 Years of Age (N = 30)**

Variable	Stand. Beta	t-value	Sig
<b>F = 17.993<sub>3,18</sub>, p = .000 (76%)</b>			
Patient Physical Functioning	.263	.960	.350
Level of Assistance Required	-.347	-1.290	.213
Mother's Perception of Child Health	.040	.361	.723
Patient Cognitive Function	.137	.829	.418
Patient Adaptive Behaviour	.539	3.292	.004**
Maternal Parent Distress	-.371	-1.477	.157
Maternal SOC Comprehensibility	.509	2.903	.009*
Maternal Distress	1.116	4.964	.000**
Family Environment: Cohesion	.717	4.524	.000**
Discrepant SOC between Parents	-.568	-3.972	.001**
Family Environment: Organisation	.473	3.605	.002**

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships illustrate how factors associated with family functioning have the biggest impact on mothers' expectations of child future achievements, when child health and maternal stress and coping are taken into consideration. This accounts for 75% of the explained variance.

Measures of adaptive behaviour have been used to define learning disability and continue to be used to assess and classify such disability. Adaptive behaviour has also been conceptualised as functional independence and theorised to mediate between the severity of disability and child adjustment (Casey et al., 2000). Indeed, in the present study adaptive behaviour shows a strong positive correlation with cognitive function (Spearman's Rho = .861,  $p < .001$ ), and is therefore a strong indicator of learning disability with this patient group. With regard to mothers' expectations of child future achievements, in the first step of the analysis, cognitive function stood out as the

single child health variable that related to expectations; and with all the other variables entered into the analysis, adaptive behaviour continues to be a significant factor. However, other factors, which relate to stress processing and social ecological factors such as mothers' coping and family functioning, also contribute to mothers' expectations. These findings will be discussed in relation to previous and future research, and to practical applications.

#### **6.4.2. Discussion**

The findings illustrate that the social-ecological factor relating to family functioning and stress processing factors relating to mothers' internal coping resources are associated with mothers' adjustment to having a child with MPS IH post-BMT, and in turn relate to her expectations of the child. By appraising the world and her situation as comprehensible, and living in a family environment that she perceives as being cohesive and organised, she is able to be more positive about her child's future achievements. The *comprehensibility* domain of SOC in particular refers to an individual's ability to see their internal and external environments as structured, predictable, and explicable. Some researchers consider the parenting of a young child with developmental disability to be one of the most stressful situations for parents (e.g. Beckman, 1991; Baxter et al., 2000). However, parents of children with developmental disability who have high SOC are reported as not suffering negative consequences as a result of the situation, since they redefine it and perceive it as a challenge (Olsson & Hwang, 2002). Aspects of the family environment have also been highlighted as being instrumental in parents' adjustment to having a child with disabilities or chronic illness (e.g. McCubbin and Patterson, 1983; Lillie, 1993; Pelchat et al., 2003), which are supported by the findings of the present study. These show positive relationships between family cohesion and organisation, and mothers' expectations. The findings suggest that parental incongruence can have a negative impact on mothers' expectations. When mothers and fathers in this sample have a different view of the world, particularly when mothers have lower SOC than fathers, it has implications for mothers' ability to be positive in terms of the child's future.

Parental risk in terms of a learned helplessness attributional style also requires attention when considering parental expectations, as a mother's tendency to lack



assertiveness, to have low self-esteem and to have low expectations of their own parental efficacy has been related to poor child outcomes in terms of their psychosocial development and cognitive function (Thompson et al., 2002). Thus low parental expectations can have a detrimental effect on the development of the child. This is possibly reflected in the findings of the present study, as mothers' expectations are positively related to child adaptive behaviour. However, this relationship could also simply illustrate mothers' tendency to be realistic in terms of her expectations when her child has more significant cognitive impairment and poorer adaptive functions. Although adaptive function has an impact on mothers' expectations of the child here, the biggest contributor to those expectations are mothers' internal coping resources and aspects of the family environment that allow her to make sense of her situation in a way that is structured and predictable. These thus buffer the effects that other child illness and functioning factors have on her appraisals of her child's condition and in turn on her expectations about the child's future. Conversely, these factors also have implications for parenting in terms of over-protection and the failure to encourage the child to develop adaptive skills and autonomous behaviour.

The relationship found between mothers' emotional distress and expectations is an anomaly however, as the relationship is positive. This does not support previous research that has linked maternal depression to negative expectations of child problems (Luoma et al., 2004). It may be possible, that while mothers can feel in control of the situation from an organised and mutually supportive family foundation, when she sees the world as structured and predictable, and when her child's learning disabilities are less pronounced, feelings of emotional distress remain. It may be possible that as a mother such feelings prevail simply as a result of the child's condition, as feelings of sadness. Despite expectations being positively related to aspects of the mother and of the family, significant physical disability and learning difficulty remain for the child. It is possible that while mothers' expectations are relatively positive (or hopeful), the child's condition remains, which is the one thing the mother cannot change. This has implications for her emotional well-being. Mothers' well-being has been associated with feelings of self-efficacy in terms of the parenting role (Pelchat et al., 2003). Part of that role is to protect the child from harm. Thus, it is possible that the presence of the MPS condition and the continued progression of bone and joint disease challenge this role.

In summary then, together with adaptive function, mothers' coping in terms of Sense of Coherence, in particular *comprehensibility* is associated with mothers' expectations of her child's future achievements. It is important then to ensure that families are provided with as much candid information as possible about long-term outcomes of BMT, in particular life expectancy. Additionally, the availability of behaviour management and educational interventions would be useful, as would contact with other families, who are either in or have been in the same situation with the same disorder. In order to promote family cohesion, it is important that both parents are involved in the decision-making process regarding any kind of support or interventions, including those pertaining to education, surgery, and the child's social activities. Family workshops, which promote a structured and joint approach to parenting may also help parents to create a more organised family environment. Workshops on parenting which focus on longer-term implications for the child may also help parents to look to the child's future, as well as living day-to-day. Longitudinal research which explores the differential ways in parents of children affected by MPS IH adjust post-BMT would be encouraged, so that more specific and appropriate support can be given to both mothers and fathers.

## **6.5. Parent Appraisals: Maternal Anxiety about Child Welfare, and Perceptions of Offspring Risk**

### **6.5.1. Mothers' Anxiety about Child Welfare**

This section of analysis explores mothers' anxiety about the welfare of their child affected by MPS IH post-BMT when participating in everyday childhood activities, and how much they consider their child to be at risk of coming to harm. An independent samples t-test was employed to explore the difference between mothers of children aged under-12 years' mean scores of anxiety ( $M = 39.87$ ,  $SD = 15.87$ ) and mothers of children aged 12-years and over ( $M = 43.54$ ,  $SD = 13.46$ ), affected by MPS IH post-BMT. Overall there was no significant difference between the mean scores, indicating that mothers of younger children did not worry any more or less about the welfare of their child carrying out everyday childhood activities than mothers of older children and young adults. However, again using an independent samples t-test, mothers of children affected by MPS IH post-BMT aged under-12 years were found to worry more ( $M = 39.74$ ,  $SD = 15.87$ ) about their children's welfare when carrying out everyday childhood activities than mothers of children *not* affected by chronic illness or disability ( $M = 31.80$ ,  $SD = 12.90$ ). This difference was found to be statistically significant ( $t = 2.399$ ,  $df = 74$ , two-tailed  $sig = .019$ ).

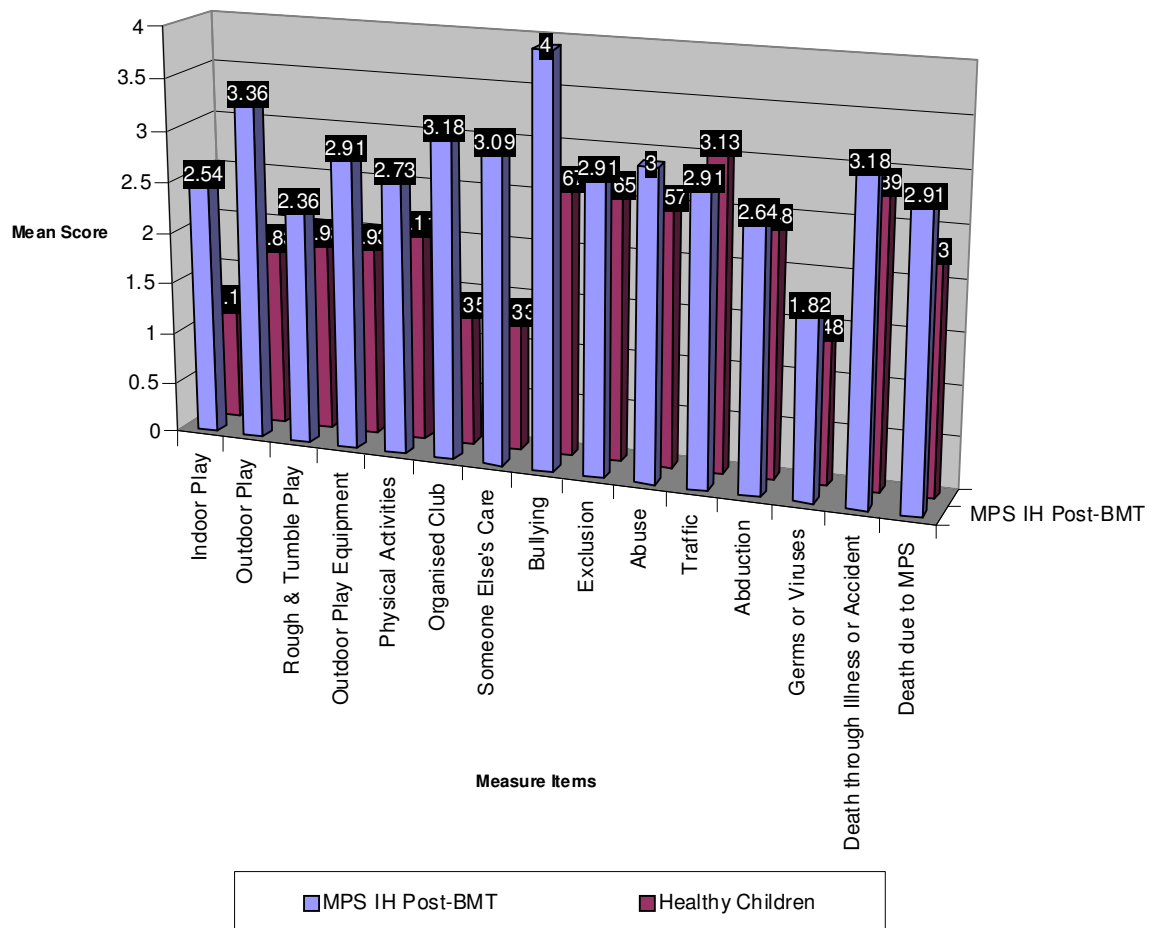
The activities the mothers of the under-12s worried the most about were the child's mortality due to the MPS condition and rough and tumble play, and the issues they worried the least about were indoor play, abduction and leaving their child in someone else's care. The implications of these findings will be discussed. Mothers of children and young people aged 12 years and over however, worried the most about the dangers of traffic. The items then highlighted as causing the most worry were accidental death, bullying, and their child being outdoors unsupervised. The activities that the mothers of the over-12s were least concerned about were germs and viruses, physical activities, and the fact that their child was potentially sexually active. Descriptive statistics for each item on the Worry Scale as completed by mothers of children and young people affected by MPS IH post-BMT are shown in Table 6-11

below. Item means for healthy and MPS IH children aged under-12 years are illustrated in Figure 6-4.

**Table 6-11 Descriptive Statistics for Mothers' Anxiety About Child Welfare**

<b>Item Under-12s (Item Over-12s)</b>	<b>Under-12s (N = 32)</b>		<b>Over-12s (N = 11)</b>	
	Mean	Std. Dev	Mean	Std. Dev
Indoor Play: Parent in Vicinity (Indoors Unsupervised)	1.56	1.56	2.54	1.75
Outdoor play: Parent in Vicinity (Outdoors Unsupervised)	2.59	1.56	3.36	1.69
Rough & Tumble Play (Potentially Sexually Active)	3.47	1.37	2.64	2.11
Outdoor Play Equipment (Evening Socialisation)	3.09	1.55	2.91	1.58
Physical Activities (Physical Activities)	2.69	1.69	2.36	1.36
Taking Child to Organised Club (Daytime Socialisation)	2.44	1.74	2.73	1.55
Someone else's care (Consented Sexual Relationship)	2.06	1.98	2.91	2.02
Being Bullied (Being Bullied)	2.87	1.79	3.18	1.83
Being Excluded by Others (Excluded)	3.03	1.82	3.09	1.45
Abuse (Abuse)	2.41	1.86	3.00	2.10
Traffic (Traffic)	3.00	1.78	4.00	1.18
Abduction (Abduction)	2.00	2.11	2.91	1.45
Germes or Viruses (Germes or Viruses)	2.72	1.76	1.82	1.66
Death through Illness or Accident (unrelated to MPS)	3.06	1.85	3.18	1.99
Natural Death (due to MPS)	3.75	1.85	2.91	1.70

**Figure 6-4. Item Means for Mothers' Anxiety about Child Welfare: Healthy and MPS IH Children Aged Under-12 years**



Although their child's mortality due to MPS was the item on the measure that the mothers of the children aged under-12 years rated as the most worrisome, and they worried more so about this ( $M = 3.75$ ,  $SD = 1.85$ ) than the mothers of the children and young people in the older group ( $M = 2.91$ ,  $SD = 1.70$ ), the difference was not found to be statistically significant between the mothers of the two age groups, using an independent samples t-test. This illustrates how mothers' perception that their child is vulnerable in terms of their mortality prevails into adulthood.

### **6.5.2. Predictive Association between Risk and Resistance Factors and Mothers' Anxiety about Child Welfare**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in mothers' anxiety about child welfare than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Illness-related risk factors that were added into the model were patient physical functioning as measured by the MPS Health Assessment Questionnaire, cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests, adaptive functioning as measured by the Vineland Adaptive Behaviour Scales, and the number of serious adverse events experienced by the child as reported by the parents in the Child Medical History Questionnaire. One intrapersonal resistance factor added into the model was maternal distress, a measure of maternal mental health as measured by the CAPI. Stress processing resistance factors that were added into the model were mothers' perception of child health, maternal SOC *comprehensibility* as measured by the Sense of Coherence Scale, maternal parenting distress as measured by the Parenting Stress Index, and maternal religious or fatalistic beliefs and attitudes as measured by the Beliefs and Attitudes Scale developed for this study. Social-ecological factors that were entered into the model were family cohesion as measured by the Child Health Questionnaire, mothers and fathers discrepant or shared views of parenting stress, and maternal feelings of disappointment regarding family members' lack of understanding regarding the child's MPS condition as measured by the Family Discord measure developed for this study. This analysis was restricted to mothers of children aged under-12 years of age as the line of questioning differed between the two age groups, and this is the larger of the two groups. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-12 below to show the relative contribution of each of the child and family dimensions to Mothers' Anxiety.

**Table 6-12 Model for Mothers' Anxiety About Child Welfare: Child Under 12 Years of Age**

Variable	Standardised Beta	t-value	Sig
<b>F = 9.137<sub>3,13</sub>, p = .002 (82%)</b>			
Patient Physical Functioning	.045	.362	.723
Mothers Perception of Child's Health	-.403	-4.069	.001**
Patient Cognitive Function	.689	2.678	.019*
Number of Serious Adverse Events	-.150	-1.196	.253
Patient Adaptive Behaviour	-.516	-2.397	.032*
Maternal Parent Distress	-1.036	-3.940	.002**
Maternal SOC Comprehensibility	-.033	-.182	.858
Maternal Distress	.586	2.520	.026*
Maternal Beliefs	.223	2.242	.043*
Discrepant Reports of Parenting Stress	.777	4.641	.000**
Disappointment: Issues Relating to Child's MPS Condition	.455	3.923	.002**
Family Cohesion	-.326	-1.617	.130

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships indicate that mothers' anxiety about her child's welfare when carrying out various everyday childhood activities is associated with two child illness/functional-related factors, cognitive and adaptive function, and to a variety of maternal intrapersonal, stress processing, and social-ecological resistance factors, which accounted for 82% of the variance. For example, mothers' mental health, her perception of child health and parenting distress, her spiritual beliefs, aspects of the family environment, in particular mothers and fathers discrepant reports of parenting stress, and immediate and extended family members' lack of understanding about the child's medical condition and the BMT. These relationships will be discussed.

### 6.5.3. Mothers' Perceptions of Offspring Risk

The Risk Perceptions questionnaire asked mothers to rate whether they believed their child to be at more, less, or equal risk of coming to harm compared to a healthy peer, when carrying out certain activities. The results show the mothers of MPS IH patients

aged 12 years and older to consider their children more at risk of coming to harm compared to a healthy peer ( $M = 57.48$ ,  $SD = 6.85$ ) than the mothers of the under-12s MPS group ( $M = 62.36$ ,  $SD = 5.73$ :  $t = -2.109$ ,  $df = 40$ ,  $p = <0.05$ ). Furthermore, in comparison to mothers of children *not* affected by chronic illness or disability ( $M = 43.07$ ,  $SD = 4.70$ ), mothers of children affected by MPS IH post-BMT felt their children to be at more risk of coming to harm compared to a healthy peer ( $M = 57.48$ ,  $SD = 6.85$ :  $t = 10.176$ ,  $df = 49$ ,  $p = .000$ ).

Mothers of children aged 12 years of age and under rated their children as being most at risk of coming to harm through rough and tumble play, playing on outdoor play equipment, being bullied, and being excluded by others. They also rated them as being at higher risk of dying naturally due to their MPS disease. These results illustrate how mothers consider their child to be at risk both physically and emotionally, and how they consider them to be vulnerable in terms of their mortality. The situations where they considered their children least at risk were indoor play, going to an organised club, and leaving the child in someone else's care.

With regard to mothers of children and young people aged 12 years and over, as with mothers of the under-12s they considered their child to be at more risk of coming to harm than another individual the same age and not affected by illness or disability on every item on the Risk Perceptions measure. Activities where mothers considered their child at most risk of coming to harm included being outside unsupervised, going out to socialise in the evening, being on the roads (traffic), and dying due to their MPS disease. These results illustrate how mothers perceive their child as vulnerable when carrying out activities associated with normal adult social independence. They also illustrate how they continue to consider their child at risk of death due to MPS IH as they grow into adulthood. The activities mothers felt their child at least risk of coming to harm included being duped into going off with someone, being vulnerable due to sexual activity potential, and germs and viruses. As with Mothers' Anxiety, these situations were considered less risky for the individual as mothers felt their child was never put in the position where they would be vulnerable to being tricked to go off with someone (as they were never out unsupervised) or likely to engage in sexual

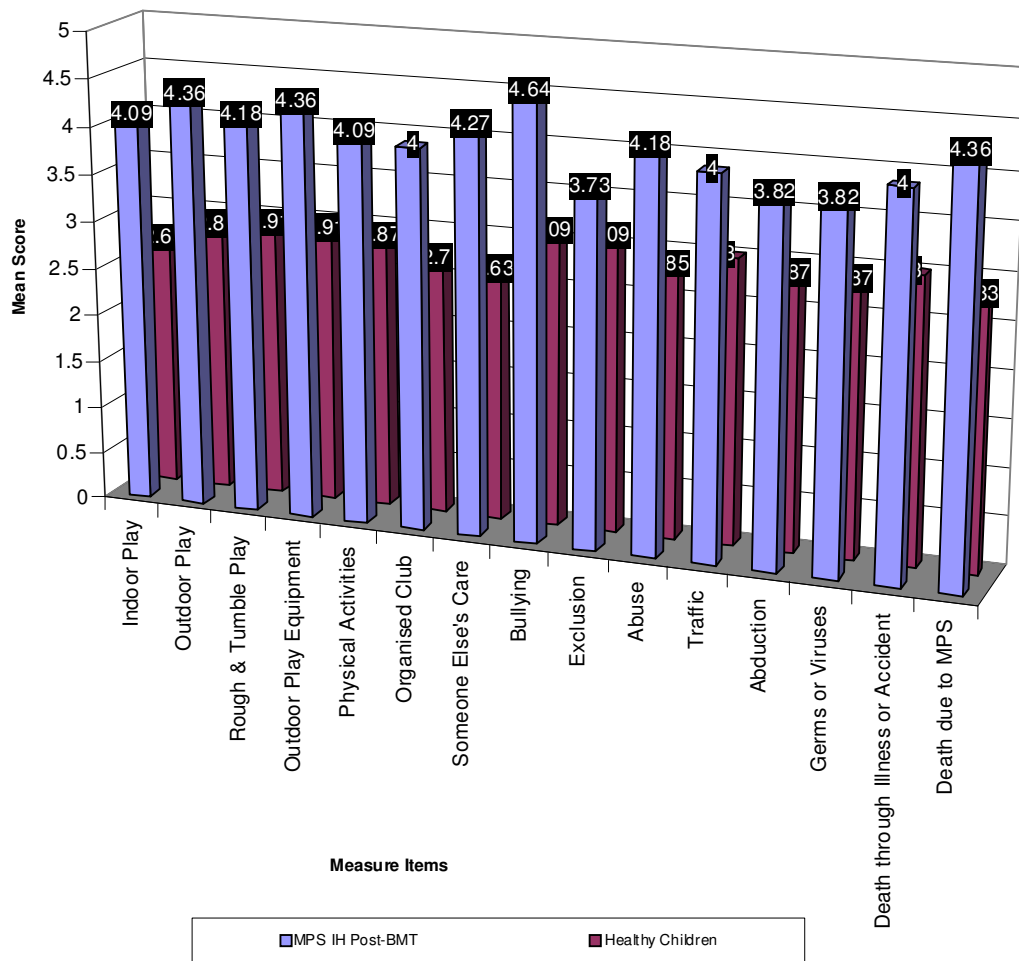


activity. Descriptive statistics for each item on the Risk Perceptions measure as completed by mothers of children affected by MPS IH post-BMT are shown in Table 6-13 below. Item means for Healthy and MPS IH children aged under-12 years are illustrated in Figure 6-5.

**Table 6-13 Descriptive Statistics for Mothers' Perceptions of Offspring Risk**

Item	Under-12s (N = 23)		Over-12s (N = 11)	
	Mean	Std. Dev	Mean	Std. Dev
Indoor play: Parent in Vicinity (Indoors Unsupervised)	3.59	.84	4.09	.83
Outdoor Play: Parent in Vicinity (Outdoors Unsupervised)	3.78	.75	4.36	.81
Rough 'n' Tumble Play (Potentially Sexually Active)	4.09	.86	3.82	.98
Outdoor Play Equipment (Evening Socialisation)	4.22	.61	4.36	.81
Physical Activities (Physical Activities)	3.91	.78	4.18	.87
Organised Club (Daytime Socialisation)	3.53	.80	4.09	.83
Someone Else's Care (Consented Sexual Relationship)	3.50	.76	4.00	.89
Being Bullied (Being Bullied)	4.09	.86	4.00	1.00
Being Excluded by Others (Exclusion)	4.03	.86	4.27	.79
Abuse (Abuse)	3.94	.84	4.18	.75
Traffic (Traffic)	3.87	1.07	4.64	.67
Abduction (Being tricked to go off with someone)	3.48	.72	3.73	.79
Germes or Viruses (Germes or Viruses)	3.81	1.03	3.82	.75
Death through Illness or Accident (unrelated to MPS)	3.66	.83	4.00	.77
Natural Death (due to MPS)	4.47	.76	4.36	.81

**Figure 6-5 Item Means for Mothers' Risk Perceptions: Healthy and MPS IH Children Aged Under-12 years**



#### **6.5.4. Predictive Association between Risk and Resistance Factors and Mothers' Perception of Offspring Risk**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in mothers' perceptions of child risk than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) psychosocial risk factors, 3) intrapersonal resistance factors, 4) stress processing resistance factors, and 5) social ecological resistance factors. Illness-related risk factors that were added into the model were patient physical functioning as measured by the MPS Health Assessment Questionnaire, cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests, and the number of serious

adverse events the child has experienced as reported by the parents in the Patient Medical History Questionnaire. Psychosocial risk factors added into the model were the level of assistance required by the child carrying out activities of daily living as measured by the MPS Health Assessment Questionnaire. One intrapersonal factor added into the model was mothers' mental health as measured by the GHQ (Goldberg and Williams, 1988). One stress processing resistance factors added into the model was mothers' perception of child health as measured by the Child Health Questionnaire. Social-ecological factors that were entered into the model were active-recreational family environment as measured by the Family Environment Scale, mothers and fathers' discrepant or shared view of the child's care needs and of parenting stress, and maternal disappointment about family members' lack of understanding about the child's MPS condition as measured by the Family Discord measure given in the parent interview. Again, this analysis was restricted to mothers of children aged under-12 years. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 6-14 below to show the relative contribution of each of the child and family dimensions to Mothers' Risk Perceptions.

**Table 6-14 Model for Mothers' Perception of Offspring Risk: Child Under-12 Years of Age**

Variable	Standardised Beta	t-value	Sig
<b>F = 8.522<sub>4,15</sub>, p = .001 (66%)</b>			
Patient Physical Functioning	.102	.685	.504
Mother's Perception of Child's Health	-.271	-1.808	.091
Patient Cognitive Function	-.195	-1.217	.243
Number of Serious Adverse Events	-.265	-1.479	.160
Mother's Mental Health	.360	1.905	.076
Family Environment: Active Recreational	.641	3.617	.003**
Discrepant Accounts of Assistance Required by Child	-.499	-3.089	.007*
Discrepant Reports of Parenting Stress	.455	2.539	.023*
Disappointment: Issues Relating To Child's MPS Condition	.328	2.381	.031*

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships indicate that stress processing and social-ecological resistance factors have the most significant impact on mothers' perception that the child is more at risk of coming to harm compared to a peer not affected by illness or disability, and they accounted for 66% of the variance. The stress processing factor relating to mothers' perception of child health demonstrates a relationship to mothers' risk perceptions. Though not a significant relationship it shows a negative trend, which indicates that mothers' perception that her child is at risk of coming to harm is greater when she perceives her child's health status to be poor. This relationship is felt to be worthy of mention in light of the small sample size. This is equally relevant for mothers' mental health. Although the relationship between mothers' risk perceptions and greater mental health problems was not significant, it shows a possible trend and is worth noting here.

With regard to the social-ecological factors of family functioning and family environment, a number of interesting relationships were found. Two measures, which were completed by both mothers and fathers, when their scores were discrepant, related to mothers' risk perceptions. In particular, they were associated when mothers experienced more parenting stress than fathers, and when fathers felt the child required more assistance with activities of daily living than the mother rated. Additionally, mothers' risk perceptions were positively related to family members', both immediate and extended, lack of understanding about the child's MPS condition and BMT treatment. This however, illustrates mothers' feelings of upset or disappointment about these issues that remain unexpressed, as opposed to verbalised feelings or disagreements. Mothers whose families were more socially active also reported greater risk perceptions. These issues will be discussed in relation to mothers' anxiety about the child's welfare and to child psychosocial outcomes.

#### **6.5.5. Discussion**

The findings of the present study illustrate how the mothers of children aged under-12 years worried the most about their child's mortality, in particular their child dying as a

direct result of MPS IH, despite the fact they had undergone BMT. The activity they rated next as causing the most worry was rough and tumble play. These findings first and foremost illustrate mothers' continued anxiety about their child's mortality, and indicate how uncertainty and concern about long-term survival post-BMT prevails. They also illustrate mothers' concern for the child coming to physical harm, and highlight the child's orthopaedic difficulties in terms of their spines, hips, and knees. With regard to length of life and whether the condition continues to be life-limiting post-BMT, there is no data available that can address this issue. In the UK there are no recorded deaths of MPS IH sufferers post-BMT, once they have survived past the crucial and risky period immediately after transplant. However, since the oldest living patient post-BMT is only 25 years of age, it will be many years before issues regarding life span can be properly addressed. Nevertheless, the uncertainty that prevails for parents regarding this issue may have wider implications for parenting style and over protectiveness, and for the child's psychosocial development. This requires further attention from medical professionals and support organisations.

With regard to concerns about physical injury that are related to the child's orthopaedic difficulties, these are realistic. Due to joint pain and stiffness, and skeletal abnormalities, children affected by MPS IH can have serious problems of this nature post-BMT. There has also been some concern about instability of the cervical spine. In other MPS diseases, particularly MPS IV such instability can be life threatening and corrective surgery is usually required (e.g. Chirossel et al., 2000; Crockard & Stevens, 1995). However, whether this is a realistic problem with MPS IH patients post-BMT is unclear. One paper has detailed the correction of odontoid dysplasia post-BMT, which reduces the potential for cervical spinal cord injury for MPS IH patients following transplantation (Hite et al., 2000); and to date, no MPS IH patients in the UK have experienced this problem post-BMT or have had corrective surgery at this point on the spine. This issue requires attention from orthopaedic surgeons who specialize in the MPS field, so that concern about spinal cord injury due to instability of the cervical spine can be minimized in this patient group through monitoring. If this issue can be removed as a concern for parents, such play in childhood would cause less worry and at least one burden would be lifted.

The issues that mothers of children aged under-12 years worried the least about were indoor play, abduction and leaving their child in someone else's care. Mothers explained that they did not worry so much about their child when they played indoors, as they would be present and supervising. They worried less about abduction because their child was never allowed to be alone and would not be put in the position where such an event could take place. Indeed, mothers rated the child as being 'less at risk' of abduction compared to a healthy peer. Regarding leaving their child with someone else, they either did not do so and therefore did not worry about it, or they only ever left their child with someone they trusted absolutely.

Mothers of children and young people aged 12 years and over worried the most about the dangers of traffic. The items then highlighted as causing the most worry were accidental death, bullying, and the individual being outdoors unsupervised. This illustrates mothers' perceived vulnerability of the child in terms of physical disability (e.g. being quick enough to cross a road, poor eyesight, etc), their mental and physical capability in terms of having the ability to look after themselves unsupervised and away from home, and their vulnerability at the hands of others. This has implications for increased independence as the child gets older and the fact that the child is more likely to partake in activities away from home and the family, where the mother perhaps has less opportunity to protect them. These are therefore activities that the mother may have less control over as the child gets older. It is worth noting here however, that the majority of the children and young people in the over-12s group did not go out unsupervised. With the exception of one individual who lived independently, and one who lived in residential care, all of the adult patient participants lived with their parents. For daytime activities such as school, college or day centre, transport was provided for all individuals except two. Those who enjoyed social activities were accompanied by either friends or family members, or were collected and dropped off by parents, with the exception of one. For the most part however, they did not partake in social activities other than family gatherings. Nevertheless mothers' expressed their concern about such activities whether the young people did them or not. It is not known at this stage whether mothers' anxiety is related to the hindrance of child independence with this patient group, but this topic warrants further investigation.

The activities that the mothers of the over-12s were least concerned about were germs and viruses, physical activities, and the fact that their child was potentially sexually active. Germs and viruses are more pertinent to younger children with MPS IH who have undergone bone marrow transplant as they are vulnerable to infection following transplant. This issue was clearly less of a concern to mothers of older children than younger children in this study. With physical activities, as the children grow older they become increasingly physically limited and partake in physical activities less and less. Those that did continue with physical activities did so in a controlled environment, for example as part of organised activities or events specially designed for individuals with disabilities, which may explain mothers' reduced concern. With regard to sexual activity potential, parents explained their lesser ratings of concern as being due to their child showing no interest in members of the opposite sex, not having the opportunity to have sexual relations, or not being mentally or physically capable of having sexual relations. Again, this issue warrants discussion and further education with this patient group, as the sexuality of young people with disabilities is often overlooked, and may hold relevance for the younger individuals in this patient group as they develop into adolescence.

Although certain issues caused mothers more anxiety and were perceived as being more risky than others, the findings of the present study demonstrate how overall the mothers of the younger children affected by MPS IH post-BMT worried more about their children's welfare and perceived them to be more at risk when carrying out everyday childhood activities than the mothers of the children not affected by chronic illness or disability. This finding would be expected, as although all parents worry about their children's welfare to a certain extent, when they are affected by physical and learning disability it is not surprising that they are deemed to be more vulnerable. Interestingly the mothers of the younger MPS children did not worry any more or any less about their children than the mothers of the adolescents and young adults. However, the older patient participants were perceived as being more at risk than the younger ones. This is possibly due to some of the items on the measure that was designed for the older patient participants pertaining to adult social independence. Compared to the behaviours presented to the mothers of the under-12s, those

presented to the mothers of the over-12s suggested more adult orientated pastimes. Whether mothers' concern about child welfare and their perceptions of risk as described in the present study can be considered to be over-concern and detrimental to the children's psychosocial development however, remains to be seen. Further research of a longitudinal nature is required to explore this topic with this patient group further.

In terms of the investigation of predictors of maternal anxiety and perceptions of risk, a number of issues that have been previously studied hold relevance here. The regression analysis in the present study indicates that mothers' anxiety about child welfare is increased when the child's adaptive skills are less proficient and cognitive function less impaired. In terms of cognitive function, outcomes of BMT can be extremely varied with some children achieving low normal intelligence while others experience severe learning difficulties. Thus, it is possible that when a child has milder learning difficulties boundaries become blurred and different issues of concern become pertinent to parents. For example, the child may wish to participate in activities with peers and feel the need for some independence from the family. However, with milder learning difficulties mothers may feel the child to have borderline coping abilities, and thus battle with wanting to allow the child the freedom to participate in peer group activities, while endeavouring to protect them from harm. This may be especially true if the child's adaptive skills are less proficient, thus rendering the child less competent in terms of communication, and social and daily living skills.

With regard to maternal intrapersonal factors, mothers' anxiety showed a positive relationship to emotional distress. This supports previous research, which demonstrates the emotional toll that can be exacted on the mother through her endeavour to ensure the child's physical and emotional well-being (e.g Anderson & Elfert, 1989; Gray, 2003; Pelchat et al., 2003). It also supports the work of Stewart and Mishel (2000) and others who have highlighted the relationship between uncertainty and psychological distress. As has been previously discussed, uncertainty regarding the child's life expectancy and other areas of the child's condition has been explored (Dodgson et al., 2000) and it has been demonstrated as impacting on



parents' psychological management of the condition (Mishel, 1983). Parents can experience uncertainty in relation to a number of areas relating to a child's condition and they can have long-term concerns about the child's future well-being (Mishel, 1981; Bonner et al., 2006). Uncertainty has also been related to a number of emotional responses including grief (Gibson, 1995), guilt and worry, and unresolved sorrow and anger (Bonner et al., 2006). Guilt and worry in particular have been positively related to perceived impact on the family and parenting worries, and it is purported that parents who score highly on this factor perceive their child as fragile (Bonner et al., 2006). In support of this, the findings of the present study also showed a relationship between maternal anxiety and mothers' perception of the child's health as poor.

Another stress processing factor, maternal beliefs in things spiritual, religious, and fatalistic was positively related to maternal anxiety about child welfare, indicating that maternal anxiety about child welfare was greater the greater her spiritual beliefs. Previous research has found people who are able to put things in a religious context to cope better than those who cannot (Venters, 1981) and religion is seen as a key resource for some parents in helping them to cope and positively appraise the illness and their situation (Hill, 1994; Williams, 1993; Kelleher and Islam, 1996). In the situation where a child has survived against all adversity, families can give special meaning to the child's place in the world and believe their survival to be fated (Mayer, 1982). It may be this however; that leads them to feeling that their child is precious and fragile, which exacerbates their anxiety about their welfare. Maternal perceptions of child vulnerability may also be linked to the relationship found between maternal anxiety about child welfare and parenting distress. Parenting distress relates to distress experienced as a direct result of the parenting experience or the parent-child system, and in the present study this was negatively related to maternal anxiety about child welfare. Indeed, if it is the mother's natural role to nurture and protect the child from harm as proposed (e.g. Bury, 1982; Young et al., 2002) and her well-being is related to the care of the child and her role as mother in their adaptation (Pelchat et al., 2003), feelings of worry about the child's welfare would not necessarily impact upon the mother's perceptions of parenting stress and would not necessarily constitute a distressing element of parenting.

The relationships found in this analysis also have implications for support within the family. The results indicate that mothers' anxiety is greater when they report greater parenting stress than fathers, and when they have unexpressed feelings of resentment towards family members (both immediate and extended) for their perceived lack of understanding about the child's condition and BMT treatment. These relationships implicate the spousal relationship and the role that a shared view of parenting may play in mothers' feelings of anxiety about the child's welfare. This supports previous research, which cites the spousal relationship (McCubbin & Patterson, 1983; Ray, 2002) and a cohesive family environment (Warfield et al., 1999) as being important to mothers' ability to adjust to having a child with illness or disability; and which finds external family members to be as much of a source of stress to mothers as they can be a source of support (e.g. Ray, 2002).

Aspects of the spousal relationship and family functioning also have implications for mothers' perceptions of risk regarding the child. The findings of the present study illustrate how the degree to which the mother perceives the child to be at risk is exacerbated when aspects of the spousal relationship, and of the wider family, are out of tune with the mother's experiences and feelings. In particular, mothers and fathers differential experiences of parenting stress, and their differing ideas about the child's needs in terms of the level of assistance the child requires carrying out activities of daily living. Mothers' unexpressed feelings of disappointment towards other family members about their lack understanding about the child's MPS condition and the parents' decision to go ahead with the BMT are also significant here. These findings illustrate how family factors might contribute to the degree to which mothers perceive their child to be at risk of coming to harm, and reinforce the similar relationship found between family factors and mothers' anxiety about child welfare. Thus, the results of this analysis demonstrate the role that resistance factors relating to intrapersonal, stress processing, and social-ecological factors play in moderating the effects that child illness and functional factors have on parent outcomes in terms of their perceptions and appraisals of child welfare and risk.

In summary then, the findings of the present study illustrate the kind of issues that mothers worry about in terms of their children's welfare and how vulnerable they

perceive them to be. They highlight the roles that the child's cognitive and adaptive functioning play in the extent to which mothers worry, and how her own internal and external coping resources can moderate this. This has implications not only for mothers' health and emotional well-being, and for the functioning of the family, but also for issues of parenting, and consequently for child psychosocial development and adjustment to living with MPS IH post-BMT. These issues therefore require attention when providing services or support to families affected by this disorder.

With regard to the issue of child death and vulnerability, it is important that families are given as clear an idea as possible about life span following BMT. Further research regarding orthopaedic difficulties and the risk of spinal cord injury would also be helpful in reducing parental (over)concern. Parenting workshops that involve both parents would also be useful by promoting a joint approach to parenting, and encouraging the development of the father-child bond. The dissemination of parenting guidelines and the implications of parenting on child psychosocial outcomes would also be useful in helping families to look to the child's future as well as living day by day. Further research of a longitudinal nature is required to explore the relationships between parental concern for child welfare and perceptions of risk, parenting style, and child psychosocial and lifestyle outcomes. Future research of this nature would also benefit from having fathers as equal participants in the surveys, so that family functioning, support, and parental attitudes can be more fully explored; and data about siblings so that their role in the reduction of maternal anxiety can be explored.

## **6.6. Summary of Findings: Parent Adjustment Outcomes**

The aims and objectives of the study relevant to this section of the analysis were to examine the moderating effects of psychosocial resistance factors on the relationship between illness-related stressors and parent adjustment, and to assess whether spouses differential experiences of parenting stress, coping, and perceptions of child care needs serve as one of those resistance factors. It was hypothesised that mothers would experience significantly more parenting stress than fathers, but that they would cope significantly better than fathers with the stresses of caring for a child affected by MPS IH post-BMT. It was also hypothesised that psychosocial resistance factors, including intrapersonal, stress processing, and social-ecological factors, would contribute to the variance in parent adjustment and that they would moderate the negative effects of illness-related stressors on adjustment outcomes.

The findings illustrate how mothers and fathers experienced similar levels of parenting stress, with a third of this parent population experiencing parenting stress to clinically significant levels. The hypothesis that mothers would report greater parenting stress than fathers was therefore not supported. Parent adjustment outcomes in terms of parenting stress and coping were however significantly predicted by psychosocial resistance factors. Mothers' parenting stress was significantly predicted by social-ecological factors pertaining to the family environment, spousal support, and a shared view of parenting (family cohesion, discrepant reports of parenting stress and of the child's care needs), accounting for 71% of the overall variance. Fathers' parenting stress was also significantly predicted by social-ecological factors pertaining to the family environment (family cohesion and discrepant reports of parenting stress), accounting for 80% of the overall variance. In terms of parent coping, fathers were found to have significantly lower SOC than mothers, particularly in the Manageability domain. This suggests that they are less able than mothers to avail themselves of the appropriate resources to enable them to cope, and supports the hypothesis that fathers will cope less well than mothers with the care of a child affected by MPS IH post-BMT. In terms of the predictors of parent coping, mothers' SOC was significantly predicted by both stress processing and social-ecological factors, which accounted for 89% of the overall variance. These pertained to mothers'

perceptions of the impact the child's illness had on the family and her perceptions of the child's overall health. They also related to family disagreements particularly in relation to family members' support and understanding regarding the child's condition. Mothers' coping was also significantly predicted by the level of assistance the child required carrying out activities of daily living, a psychosocial risk factor. Fathers' SOC was significantly predicted by stress processing and social-ecological factors that were associated with the family environment (discrepant reports of parenting stress and family disagreements relating to the mother's role) and to perceptions of the impact the child's condition had on the family, which accounted for 57% of the variance.

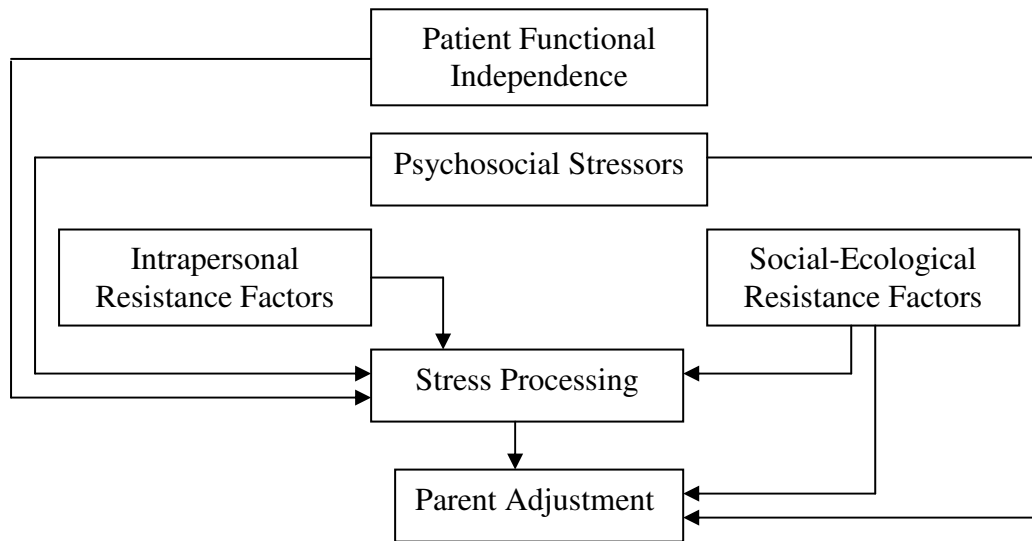
In relation to mothers' mental health, a number of stress processing and social-ecological variables showed significant relationships. These included mothers' perceptions of child health, maternal parenting stress and coping (SOC), aspects of the family environment (mothers and fathers discrepant reports of parenting stress), and social support. A psychosocial risk factor also significantly predicted this outcome, which was the number of visits the child made in the previous 12 months to the MPS specialist hospital. These variables accounted for 71% of the variance in mothers' mental health. Maternal expectations of child future achievements were significantly predicted by intrapersonal, stress processing, and social-ecological factors. These were related to maternal distress, maternal coping (SOC *comprehensibility*), family cohesion and an organised family environment, and discrepant reports of parenting stress between mothers and fathers. Mothers' expectations were also impacted upon by the child's adaptive behaviour. Thus, although functional independence was an illness-related risk factor that significantly predicted maternal expectations of child future achievements, resistance factors also contributed to the variance. These variables accounted for 76% of the variance in this outcome. Similarly, mothers' anxiety about the child's welfare was significantly predicted by a number of intrapersonal, stress processing and social-ecological factors pertaining to maternal distress, maternal perceptions of the child's overall health, maternal parenting distress, religious or fatalistic beliefs, and the support and understanding of the family, they were also significantly predicted by illness-related risk factors, namely cognitive and adaptive functioning. These variables accounted for 82% of the overall variance in mothers' anxiety about child welfare. Mothers' perceptions of child risk

however, were predicted by social-ecological factors pertaining to the family environment only, accounting for 66% of the overall variance.

Overall then, although child illness-related factors associated with functional independence were shown to impact upon two of these parent adjustment outcomes, intrapersonal, stress processing, and social-ecological factors were also found to significantly contribute. The hypothesis that psychosocial resistance factors would significantly contribute to the variability in parent adjustment to having a child affected by MPS IH post-BMT, and that they would moderate the negative effects of illness-related stressors on adjustment was therefore supported. These findings also support the hypothesis that discrepant accounts of parenting stress and of the child's care needs between mothers and fathers would serve as a social-ecological resistance factors and significantly contribute to the variance in parent adjustment. The hypothesis that parents' discrepant reports regarding their coping abilities would do the same was not supported however. These findings demonstrate how biomedical risk factors pertaining to functional independence impact on parent adjustment in terms of stress processing, particularly in areas that relate to maternal perceptions of child vulnerability. They also demonstrate however, how the relationship between illness-related stressors and parent adjustment is moderated by psychosocial resistance factors. These relationships are illustrated in Figure 6-6 overleaf. The model supports the risk-resistance framework of adjustment to chronic illness proposed by Wallander and colleagues (e.g. 1989; 1992) though it does not indicate a direct relationship between patient functional independence and classic parent adjustment outcomes, namely mothers' mental health.

Coping abilities, family support and functioning, and appraisals of the child's health and abilities, and of the parents' situation have huge implications for parent adjustment when they have a child affected by MPS IH post-BMT. Since the majority of the parent adjustment outcomes investigated here relate to stress processing, these factors have implications for a number of other parent adjustment outcomes. They also have implications for parenting style and in turn child stress processing, coping, and adjustment. This is illustrated in the analysis of the patient participant adjustment outcomes and is discussed in the following chapter.

**Figure 6-6. Model for Parent Adjustment**



## CHAPTER SEVEN

### RESULTS

#### PATIENT ADJUSTMENT OUTCOMES

#### **7.0 Patient Adjustment Outcomes**

This chapter presents the findings of this study which pertain to patient participant adjustment outcomes. Findings are presented in five sections and examine patient social competency, personal adjustment, cognitive and adaptive function, internalising and externalising behaviour, and self-image. Data are examined in relation to normative samples and patient age. Patient participant outcomes are also examined using the disability stress-coping model. Thus, the moderating effect of psychosocial resistance factors (intrapersonal, stress processing, and social-ecological) on the relationship between biomedical risk factors and patient adaptation is investigated.

#### **7.1. Patient Social Competency**

The results presented in this section were presented in a paper that was accepted for publication in the journal *Child: Care, Health, and Development* on 30<sup>th</sup> September 2008. They describe the social competency of this patient population by examining the data collected from mothers when completing the Patient Socialisation questionnaire designed for this study and three subscales from the Behaviour Assessment System for Children Parent Form (Reynolds and Kamphaus, 1998): Withdrawal, Adaptive, and Social Skills. A copy of the paper accepted for publication can be found in Appendix Q.



### 7.1.1. Patient Socialisation

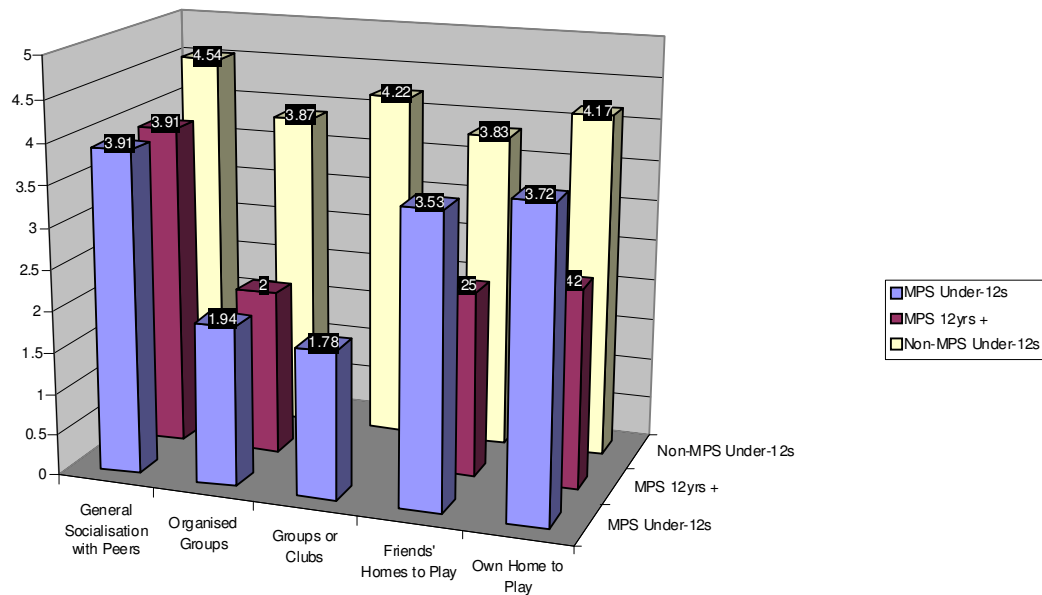
As well as asking mothers of children affected by MPS IH post-BMT to complete the Socialisation questionnaire, mothers of children aged under-12 years *not* affected by disability or chronic illness were also asked to complete this measure, as detailed in section 5.6.4. of the previous chapter. Table 7-1 below shows the number, range and mean age of participant in each of the groups.

**Table 7-1 Number, Range, and Mean Age of Participant in Each Group**

Age Group	MPS Group				Normative Group			
	Min	Max	Mean	N	Min	Max	Mean	N
Under-12	1	11	5.6	32	3	11	6.7	46
12 years +	13	25	20.6	12				

A one-way ANOVA was employed to explore the differences between the mean total scores of socialisation for children aged under-12 years affected by MPS IH post-BMT ( $M = 14.87$ ,  $SD = 5.32$ ), individuals aged 12 years of age and over affected by MPS IH post-BMT ( $M = 13.25$ ,  $SD = 6.62$ ), and children not affected by any disability or chronic condition ( $M = 20.63$ ,  $SD = 3.25$ ). The difference was found to be significantly different ( $F = 21.013$ ,  $p < .001$ ). Using the Tukey HSD multiple comparison test to explore the differences between the groups, the normative group were found to generally socialise with peers more frequently than both the MPS IH groups ( $p < .001$ ). Figure 7-1 below shows the mean scores for each item of the Socialisation scale for the three groups.

**Figure 7-1. Item Means for Socialisation Scale by Group**



As illustrated in Figure 7.1, the children not affected by disability or chronic illness scored more highly than both MPS IH groups on each item of the Socialisation measure. The significance of the differences on each item is explored using the Mann-Whitney non-parametric t-test applying the Bonferroni adjustment. Thus the required alpha level is  $p = .01$ . A separate test was conducted between the two age groups affected by MPS IH (under- and over-12 years); and between the normative sample of under-12s and the under-12s affected by MPS IH post-BMT for each item:

Item 1: *Outside of school/nursery my child gets to play with children his/her own age.* The under-12s in the normative sample ( $M = 4.54$ ,  $SD = .69$ ) were found to socialise with peers outside of school or nursery significantly more frequently than the children aged under-12 years affected by MPS IH post-BMT ( $M = 3.91$ ,  $SD = 1.42$ ) ( $U = 62$ ,  $p = .047$ ). No significant differences were evident between the under- and over-12s affected by MPS IH post-BMT on this item.

Item 2: *Outside of school/nursery I take my child to organised social groups such as play groups, toddler groups, brownies, rainbows, cubs, scouts.* The under-12s in the normative sample were found to participate in organised group activities significantly more frequently ( $M = 3.87$ ,  $SD = 1.48$ ) than the children aged under-12 years affected

by MPS IH post-BMT ( $M = 1.94$ ,  $SD = 1.68$ ) ( $U = 57$ ,  $p < .001$ ). There was no statistically significant difference between the under- and over-12s affected by MPS IH post-BMT on this item.

Item 3: *Outside of nursery/school my child attends groups or clubs (i.e. swimming, drama, music).* The under-12s in the normative sample were found to participate in club activities significantly more frequently ( $M = 4.22$ ,  $SD = 1.17$ ) than the under-12s in the MPS IH post-BMT group ( $M = 1.78$ ,  $SD = 1.45$ ) ( $U = 89$ ,  $p < .001$ ). Regarding individuals affected by MPS IH post-BMT aged 12-years and over, this item asked how often they holidayed without parents, including holidays tailored for children with disabilities. Thus, it did not make theoretical sense to perform a statistical comparison. However, twenty-seven percent participated in such activities regularly (1-2 times per year), while 55% did so rarely or never.

Item 4: *Outside of nursery/school I specifically take my child to friends' houses to play.* The under-12s in the normative sample were not found to participate in social activities at friends' houses any more or less frequently ( $M = 3.83$ ,  $SD = 1.04$ ) than the children aged under-12 years in the MPS IH post-BMT group ( $M = 3.53$ ,  $SD = 1.68$ ). However, of the participants affected by MPS IH, those aged under-12 years were found to socialise more frequently at friends' houses than those aged over-12 years ( $M = 2.25$ ,  $SD = 1.60$ ). This difference was found to be statistically significant ( $U=85$ ,  $p=.01$ ). Thus, observations of the means for the groups on this item suggest that, of the three groups, those affected by MPS IH post-BMT aged 12 years and over socialised the least with friends at friends' homes.

Item 5: *Outside of nursery/school my child has friends over to play.* The under-12s in the normative sample were not found to socialise with peers within their own homes any more or less frequently ( $M = 4.17$ ,  $SD = .80$ ) than the under-12s affected by MPS IH post-BMT ( $M = 3.72$ ,  $SD = 1.49$ ). However, of the participants affected by MPS IH, those aged under-12 years were found to do so more frequently than those aged 12-years and over ( $M = 2.25$ ,  $SD = 1.42$ ). This difference was found to be significant ( $U = 81$ ,  $p=.007$ ). Thus, again, observations of the means for the groups on this item suggest that, of the three groups, those affected by MPS IH post-BMT aged 12 years and over had friends over to socialise at their own homes the least.

### **7.1.2. Patient Withdrawal, Adaptive and Social Skills**

Patient withdrawal, adaptive and social skills were explored using the three age parameters of the BASC measure. These were 2<sup>1</sup>/<sub>2</sub>-5 years, 6-11 years, and 12-18 years. The following section describes the scores of 39 MPS IH patients post-BMT. Due to the age parameters of the measure five participants were too young to be administered the measure, and thus have been omitted from this element of the analysis. Normative scores discussed in this section refer to those set out in the BASC manual and not to the normative sample previously discussed in this section.

The children aged 6-11 years have the highest mean adaptive skills scores (M = 45.67, SD = 7.84) compared to those aged 2<sup>1</sup>/<sub>2</sub> to 5 years (M = 35.83, SD = 8.84) and 12 years plus (M = 38.83, SD (12.92). The normative mean for each of the BASC scales and composite scores is 50 with a standard deviation of 10. In comparison to normative data, Table 7-2 shows the mean scores for each of the composite scores. With regard to the subscale for Adaptive Skills, the scores for this patient group appear to be low, especially for the youngest and oldest age groups. Scores below 40 on this composite measure indicate low adaptive skills.

**Table 7-2: Descriptive Statistics for Each of the BASC Subscales across the Age Groups**

BASC Sub-Scale	Age Group	N	Min	Max	Mean	Std. Dev.
Child Adaptive Skills	2 ½ to 5 years	12	24	54	35.8	8.84
	6-11 years	15	26	58	45.7	7.84
	12 years plus	12	15	61	38.8	12.92
	Total	39	15	61	40.5	10.58
Withdrawal*	2 ½ to 5 years	12	35	75	57.3	11.93
	6-11 years	15	35	95	51.9	15.48
	12 years plus	12	37	97	59.2	19.21
	Total	39	35	97	55.8	15.71
Social Skills	2 ½ to 5 years	12	23	57	34.6	10.65
	6-11 years	15	31	60	50.8	7.42
	12 years plus	12	17	60	42.8	13.08

\* NB on the Withdrawal subscale a lower score is indication of better adaptation.

In order to test the significance of these observed differences a one-way MANOVA was performed and a Bonferroni adjustment applied. Thus the required alpha level for 95% confidence was  $p = .017$ . With respect to the relationship between the age of the child and BASC Adaptive Skills scores, a borderline significant difference was found ( $F = 3.517$ ,  $p = .040$ ). Using the Tukey HSD multiple comparison test to explore the differences between child Adaptive Skills scores at the three levels of age, children in the 6-11 years age group scored significantly higher than the children in the 2½ to 5 years age group ( $p = .039$ ). Thus, the adaptive skills of this patient group, as measured by the Behaviour Assessment System for Children, appear to be adequate for children aged 6-11 years, but low for those aged 5 years and under and those aged 12 years and over.

On examining the BASC Withdrawal scale a similar pattern emerges. The mean scores obtained by children aged 6-11 years ( $M = 51.87$ ,  $SD = 15.48$ ) are lower than those obtained by children aged 5 years and under ( $M = 57.33$ ,  $SD = 11.93$ ) and

children aged 12 years and over ( $M = 59.25$ ,  $SD = 19.21$ ), as shown in Table 7.2 above. However, unlike the Adaptive Skills scores, the mean scores for Withdrawal are within the normal range, although the mean score for the oldest group is nearing a level of withdrawal that would be of concern. Whilst this trend is not statistically significant ( $F = .808$ ,  $p = .454$ ), the partial eta squared value indicates that the observed differences ( $\eta^2 = .04$ ) demonstrate a small to moderate effect size and thus warrant further investigation.

With regard to BASC Social Skills, the same pattern emerges between the age groups. Again, the 6-11 age group's mean score is higher ( $M = 50.80$ ,  $SD = 7.42$ ) than that of the 2<sup>1</sup>/<sub>2</sub> to 5 year age group ( $M = 34.58$ ,  $SD = 10.65$ ) and that of the 12 years plus age group ( $M = 42.83$ ,  $SD = 13.08$ ), as shown in Table 7.2 above. Thus, the children in the 6-11 years and 12 years plus age groups appear to have developed adequate Social Skills, although the 12 years plus age group's mean score is at the lower end of the normal range. Of particular concern however, is the youngest age group mean score on the Social Skills scale, which is below 40. This signifies that the children's social skills are significantly delayed in this age group and that they are in need of some social skills training.

The results demonstrate a significant overall effect for child's age on BASC Social Skills scores ( $F = 8.112$ ,  $p = .001$ ). Using the Tukey HSD multiple comparison test to explore the differences between child Social Skills scores at the three levels of age, children in the 6-11 years age group scored significantly higher than the children in the 2<sup>1</sup>/<sub>2</sub> to 5 age group ( $p = .001$ ).

### **7.1.3. Predictive Association between Risk and Resistance Factors and Patient Withdrawal, Adaptive and Social Skills**

#### **7.1.3.1. Patient Adaptive Skills**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient adaptive skills than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Risk factors that were added into the model were patient physical functioning and level of assistance required as measured by the MPS Health Assessment Questionnaire, cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests, and adaptive functioning as measured by the Vineland Adaptive Behaviour Scales. Intrapersonal resistance factor added into the model were internalising problems as measured by the BASC Parent Form and patient self-esteem as measured by the Child Health Questionnaire. Stress processing resistance factors that were added into the model were maternal parenting stress as measured by the Parenting Stress Index, maternal SOC *comprehensibility* as measured by the Sense of Coherence Scale, maternal mental health as measured by the GHQ, two measures of mothers' perception of child health as measured by the Child Health Questionnaire, and mothers' anxiety about child welfare as measured by the anxiety and risk perceptions measure designed for this study. Social-ecological factors that were entered into the model were family cohesion as measured by the Child Health Questionnaire, independent and active-recreational family environment as measured by the Family Environment Scale, and mothers and fathers discrepant or shared coping abilities as measured by the Sense of Coherence Scale. The standardised regression coefficients, t-values, and probabilities for the final model is reported in Table 7-3 below to show the relative contribution of each of the child and family dimensions to patient adaptive skills.

**Table 7-3 Model for Patient Adaptive Skills**

Variable	Standardised Beta	t-value	Sig
<b>F = 3.332<sub>4,10</sub>, P = .056 (78%)</b>			
Patient Physical Functioning	-.210	-.858	.411
Level of Assistance Required	.382	1.309	.220
Mother's Perception of Child's Health	.158	.754	.468
Patient Cognitive Function	.147	.460	.655
Patient Self Esteem	.331	2.657	.024*
Patient Internalising Problems	-.346	-1.376	.199
Patient Adaptive Behaviour	.345	1.067	.311
Maternal Parenting Stress	-.462	-2.093	.063
Maternal SOC Comprehensibility	-.576	-2.507	.031*
Maternal Mental Health	.101	.538	.602
Mother's Perception of Child Health 2	-.065	-.418	.685
Mother's Anxiety about Child Welfare	-.314	-2.185	.054
Family Cohesion	0.272	-1.473	.172
Family Environment: Independence	.422	2.704	.022*
Family Environment: Active Recreational	.344	2.228	.050*
Discrepant SOC Scores between Parents	.052	.335	.744

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships indicate that intrapersonal, stress processing and social-ecological resistance factors have the biggest impact on patient adaptive skills, accounting for 78% of the variance. The intrapersonal factor of self-esteem showed a positive relationship to patient adaptive skills, as did two social-ecological factors relating to family environment, having an independence- and active-recreational orientated family environment. One stress processing factor relating maternal sense of coherence *comprehensibility* was also related to patient adaptive skills, but showed a negative relationship, and of borderline significance was the negative relationship between mothers' anxiety about child welfare, another stress processing factor, and patient adaptive skills. These relationships will be discussed.



#### **7.1.3.2. Patient Withdrawal**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient withdrawal than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Risk factors that were added into the model were patient physical functioning and level of assistance required as measured by the MPS Health Assessment Questionnaire, and cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests. Intrapersonal resistance factors added into the model were patient self-esteem as measured by the Child Health Questionnaire, adaptive skills as measured by the BASC Parent Form, and patient mental health as measured by the Child Health Questionnaire. Stress processing resistance factors that were added into the model were maternal parenting stress as measured by the Parenting Stress Index, maternal SOC as measured by the Sense of Coherence Scale, two measures of mothers' perception of child health as measured by the Child Health Questionnaire, and mothers' anxiety about child welfare as measured by the anxiety and risk perceptions measure designed for this study. Social-ecological factors that were entered into the model were family cohesion as measured by the Child Health Questionnaire, independent and active-recreational family environment as measured by the Family Environment Scale, and mothers and fathers discrepant or shared views of parenting stress as measured by the Parenting Stress Index. The standardised regression coefficients, t-values, and probabilities for the final model is reported in Table 7-4 below to show the relative contribution of each of the child and family dimensions to patient withdrawal.

**Table 7-4 Model for Patient Withdrawal**

Variable	Standardised Beta	t-value	Sig
<b>F = 3.270<sub>4,10</sub>, P = .059 (77%)</b>			
Patient Physical Functioning	.177	.607	.558
Level of Assistance Required	-.035	-.105	.918
Mother's Perception of Child's Health	-.134	-.622	.548
Patient Cognitive Function	.116	.850	.415
Patient Self Esteem	.083	.564	.585
Patient Mental Health	-.188	-1.241	.243
Patient Adaptive Skills	-.512	-2.275	.046*
Maternal Sense of Coherence	-1.024	-3.766	.004**
Maternal Parenting Stress	-.340	-1.399	.192
Maternal Anxiety about Child Welfare	.056	.301	.770
Maternal Perceptions of Child Health 2	.223	1.119	.289
Family Cohesion	.483	2.738	.021*
Family Environment: Independence	.067	.375	.716
Family Environment: Active Recreational	-.029	-.150	.883
Discrepant Reports of Parenting Stress	-.202	-1.102	.296

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships indicate that intrapersonal, stress processing and social-ecological resistance factors have the biggest impact on patient withdrawal, accounting for 77% of the variance. The intrapersonal factor of adaptive skills showed a negative relationship to patient withdrawal, as did the stress processing factor of maternal coping (SOC). The social-ecological factor of an independence orientated family environment showed a positive relationship to patient withdrawal. These relationships will be discussed.

### **7.1.3.3. Patient Social Skills**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient social skills than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Risk factors that were added into the model were patient physical functioning and level of assistance required as measured by the MPS Health Assessment Questionnaire, and cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests. Intrapersonal resistance factors added into the model were patient self-esteem as measured by the Child Health Questionnaire, patient mental health as measured by the Child Health Questionnaire, and internalising problems as measured by the BASC Parent Form. Stress processing resistance factors that were added into the model were maternal parenting stress as measured by the Parenting Stress Index, maternal SOC *comprehensibility* as measured by the Sense of Coherence Scale, maternal mental health as measured by the GHQ, two measures of mothers' perception of child health as measured by the Child Health Questionnaire, and mothers' anxiety about child welfare as measured by the anxiety and risk perceptions measure designed for this study. Social-ecological factors that were entered into the model were family cohesion as measured by the Child Health Questionnaire, independent and active-recreational family environment as measured by the Family Environment Scale, and mothers and fathers discrepant or shared coping abilities as measured by the Sense of Coherence Scale. Again, the standardised regression coefficients, t-values, and probabilities for the final model is reported in Table 7-5 below to show the relative contribution of each of the child and family dimensions to patient social skills.

**Table 7-5 Model for Patient Social Skills**

Variable	Standardised Beta	t-value	Sig
<b>F = 8.481<sub>4,10</sub>, P = .003 (86%)</b>			
Patient Physical Functioning	.003	.015	.988
Level of Assistance Required	.319	1.288	.227
Mother's Perception of Child's Health	.092	.553	.592
Patient Cognitive Function	.692	3.634	.005*
Patient Self Esteem	.249	2.390	.038*
Patient Mental Health	.007	.040	.969
Patient Internalising Problems	-.600	-1.998	.074
Maternal Parenting Stress	-.490	-2.375	.039*
Maternal SOC Comprehensibility	-.911	-4.360	.001**
Maternal Mental Health	.017	.119	.908
Maternal Perceptions of Child Health 2	-.037	-.293	.775
Maternal Anxiety about Child Welfare	-.184	-1.701	.120
Family Cohesion	-.221	-1.670	.126
Family Environment: Independence	.464	3.612	.005**
Family Environment: Active-Recreational	.446	3.933	.003**
Discrepant SOC Scores between Parents	.035	.275	.789

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships indicate that patient functioning, intrapersonal, stress processing and social-ecological resistance factors appear to have the strongest association with patient social skills, accounting for 86% of the variance. The risk factor of patient cognitive function showed a positive relationship to patient social skills, as did the intrapersonal factors of self-esteem and patient internalising problems, though the latter relationship was of borderline significance. The stress processing factor of maternal parenting stress showed a negative relationship to patient social skills, as did maternal coping (SOC). The social-ecological factors relating to family environment also showed a positive relationship to patient social skills, particularly an independence orientated and active-recreational orientated family environment. These relationships will be discussed.

#### **7.1.4. Discussion**

The aim of this section of analysis was to explore the frequency with which MPS IH patients socialised with peers and the type of activities they took part in post-BMT. The scores of the under-12 MPS group were compared with those of a normative sample of the same age. Additionally, two groups of MPS IH post-BMT patients were compared, those under-12 years of age and those 12 years of age and over. This section of analysis also aimed to investigate patients' scores of withdrawal, adaptive and social skills in terms of patient age group as set out by the BASC measure, and in comparison to the norms of the measure. The findings illustrate how children and young people affected by MPS IH post-BMT have significantly different social experiences to those not affected by disability or chronic ill health.

Considering children aged under-12 years, those not affected by disability socialised generally with their peers outside of school or nursery more frequently than those affected by MPS IH post-BMT; they attended organised social groups; and participated in extra curricular activities such as music, sport, or swimming clubs more frequently. However, they did not socialise with peers within the home, a friend's home or the participant's home, any more frequently than children affected by MPS IH post-BMT. Considering the young people affected by MPS IH post-BMT in adolescence and early adulthood, they were not found to participate in social activities overall any more or less frequently than the children affected by MPS IH post-BMT aged under-12 years. Like the under-12s, they rarely participated in group activities. However, the older group of MPS patients did socialise with friends both within the home and at friends' homes significantly less frequently than the group of MPS IH children aged under-12 years. Therefore, for the most part, the majority of the social activities in which the children affected by MPS IH post-BMT aged under-12 years participated took place either at their own home or within someone else's home. For the over-12s, since they rarely participated in such social activities, it appears that outside of school or college, very few social activities with peers were participated in at all.

Overall the children and young people in this study group have adaptive skills that are not adequately developed. This is particularly the case for children aged 2½ to 5 years and for children and young people aged 12 years and over. In terms of withdrawal the children and young people scored within the normal range as a group, however the oldest and the youngest groups' mean scores edged towards a level that would be of concern. This pattern is similar for social skills, with the youngest children's group mean score being the lowest. These findings therefore suggest the children aged 6-11 years to have developed adequate adaptive and social skills and to not experience withdrawal, while the younger and older children in this sample have difficulties in these areas. These three characteristics are essential to an individual's social competency and to their ability to function in the world.

With regard to the younger children, aged 2½ to 5 years, it is not surprising that they may be withdrawn and timid in social situations and to lack certain adaptive skills. For this patient group, their early infant years are dominated by ill health and numerous hospital visits, referrals, examinations, tests, and sometimes surgery. This is then followed by a bone marrow transplant, a serious and invasive procedure, which renders the child extremely unwell. Subsequently the child is kept in semi isolation for up to a year following BMT. Thus, social experiences are perhaps not the norm for these children in their early years. They will have spent the majority of their time with a few select members of the family, in particular the mother, and would likely be quite delayed in the development of their social skills. Ill health and pain would also likely render a young child somewhat withdrawn for a time, until recovery and reintegration has taken place post-BMT. Few studies have explored the adaptation of infant recipients following bone marrow transplant. However, research indicates BMT to impact on children psychologically, reporting them as being more stressed and having low social competence, low self-esteem, emotional difficulties, and multiple concerns upon discharge (e.g. Wiley and House, 1988; McConville et al., 1990; Phipps et al., 1995). Children have also been reported as experiencing post-traumatic stress symptoms post-BMT (Pot-Mees and Zeitlin, 1987; Pot-Mees, 1989; Stuber et al., 1991). No such studies have explored very young infants' adaptation post-BMT who are affected by MPS or related diseases.

For the present patient population adaptive and social skills appear to be adequately developed in the children aged 6-11 years. They also do not appear to be withdrawn. However, for the children and young people aged 12 years and above, these skills are not adequately developed, and withdrawal appears to again near a level that would be of concern. Previous research has indicated a transitional point in middle childhood around the time of puberty with regards to young people's social adaptation. For example, children affected by chronic illness or disability have been found to adapt well emotionally and socially (e.g. Ievers & Drotar, 1996), whereas adolescents are found to experience delays in their psychosocial development (McAnarney, 1985) and to exhibit difficulties in their social and peer relationships (e.g. Frankel, 1996; Pihl & McLarnon, 1984; Skar, 2003). These disparate findings may be indicative of the emergence in middle childhood of subtle personality traits that are temporarily adaptive to observable disability, which then manifest as emotional, behavioural, and social difficulties in later years. This suggests that psychosocial functioning in adolescence remains similar to such functioning in early childhood, rather than changing as a consequence of their growing maturity (Pless & Pinkerton, 1975). In light of this finding it is important that this patient population is studied longitudinally, so that such nuances of development and adaptation to disability can be examined in relation to outcome measures and more importantly to give direction for quality of life enhancing initiatives.

These important points notwithstanding, the disability stress-coping model highlights the role that resistance factors play in buffering the effects that illness and disability related stressors have on adjustment. This argument is supported by the present study, as intrapersonal, stress processing, and social-ecological factors emerged as more significant contributors to patient adaptive and social skills than disability- or illness-related factors alone. In terms of adaptive and social skills, the intrapersonal resistance factor of self-esteem stood out as a significant contributor, as did having a lower incidence of internalising problems. The relationship of this latter variable with patient social skills was however of borderline significance. Previous research has highlighted self-esteem as having a significant impact on the adaptation of chronically ill children and adolescents (Anderson et al., 1982; Burlew et al., 2000; Thompson et al., 1993; Wallander et al., 1989) as has a sense of adaptive competence (Hurtig and

White, 1986). Greater social assertiveness and higher levels of self-esteem have also been associated with lower levels of anxiety and depression amongst chronically ill adolescents (Burlew et al., 2000).

The remainder of variables that impacted on child adaptive skills consisted of parent stress processing and social ecological factors. Previous research has highlighted family stress processing and other resistance factors as functioning as stress processing factors relevant to children's adaptation (e.g. Burlew et al., 2000). Family factors in particular are purported to either exacerbate or attenuate the impact the disease has on the individual (Stuber, 1996; Ostroff et al., 2000), and to be related to the development of a child's self esteem, coping resources, and overall child adjustment (e.g. Antle, 2003; Burlew et al., 2000; Wallander et al., 1989). In the present study two family functioning factors, independence and active-recreational orientation emerged as significant contributors. This was also the case for patient social skills. This is supported by previous research, which has found independence and recreational orientation in families to be associated with better adolescent adaptation to living with chronic illness, especially in terms of social competence (Moos and Moos, 2002). Thus, self-esteem, which in itself is an adaptive trait, stands out as a contributor to child adaptive skills in this patient group, along with a family environment that encourages independence and has the tendency to seek out active recreational pursuits.

The stress processing factors that stood out as contributors to patient adaptive and social skills were maternal parenting stress and maternal coping in the form of the *comprehensibility* dimension of the Sense of Coherence Scale (SOC). For patient adaptive skills maternal anxiety about child welfare also stood out as a contributor. Low maternal parenting stress and low maternal anxiety about the child's welfare contributed to the development of better patient adaptive skills. These relationships are indicative of mothers' perceptions of her parenting role and the parent-child system, and of maternal perceptions of child vulnerability. Previous research has related maternal distress to child psychosocial maladjustment (Lavigne and Routman, 1993; Thompson et al., 2002) and maternal over-involvement with and over-protectiveness of the child to behavioural and emotional difficulties (Cappelli et al., 1989; Burbach et al., 1989; Holmbeck et al., 2002). Parental anxiety and perceptions



of child vulnerability in particular have been related to child internalising problems (Estroff et al., 1994), increased social anxiety in children (Anthony et al., 2003), and illness-related anxiety and the development of anxious attachments to parents (Odegard, 2005).

With regard to maternal SOC *comprehensibility* however, this relationship is difficult to interpret, as one would expect the healthy development of patient adaptive and social skills to have a positive relationship with maternal SOC. However, in the present study, negative relationships emerged between this variable and both of these patient outcome measures. This suggests that the less structured, predictable, and explicable the mother finds the world, the better developed the child's adaptive and social skills. It is possible that this result indicates a maternal attitude that focuses on the present rather than the future, one that prioritises the child's quality of life and healthy adjustment, despite living in an unpredictable and incomprehensible situation. Thus, the emphasis for the mother is perhaps on the child's enjoyment of life however long or short it may be. It may also be possible that when the mother's SOC is low, the child learns to be less reliant on her and is forced to develop better adaptive skills. Alternatively, it may be possible that, though mothers feel their situation to be unpredictable and inexplicable, in their role as mother they protect the child from the reality of the situation and continue to endeavour to equip the child with the skills of life. Although unable to have a clear picture of the child's future, these results suggest mothers who focus on the child's adaptation with a positive attitude that encourages independence and participation in the world, a strong sense of self is developed and along with it adequate adaptive and social skills that facilitate social competence in the individual.

In addition to the intrapersonal, and parent stress processing and social ecological factors discussed, patient cognitive function emerged as having an impact on patient social skills, with better social skills being related to higher cognitive function. Again, issues relating to learning disability, academic failure, and learned helplessness may be pertinent here, as children and young people with learning disability have been found to experience social problems, in terms of withdrawal and inhibition, as a result of their self-esteem and self-concept being negatively impacted upon by academic failure (Hersh et al., 1996). Better cognitive function also indicates more proficient

communication, social, and adaptive skills (Sparrow et al., 1984). Indeed, these findings are further supported by the present study, as patient withdrawal is negatively related to patient adaptive skills. The importance of adaptive competencies has been highlighted in normally developing children (Cicchetti and Sparrow, 1990; Luthar, Woolston, Sparrow, and Zimmerman, 1995) and the lack of developmental competencies illuminated as a risk factor associated with maladjustment amongst those with chronic illness (e.g. Casey et al., 2000).

When considering patient withdrawal, parent stress processing and social ecological factors again show some relation, through in particular maternal SOC and family cohesion. Contrary to the relationship found between maternal SOC *comprehensibility* and patient adaptive skills in the present study however, maternal SOC is negatively related to patient withdrawal in this instance. Thus, in support of previous research that relates maternal emotional well-being and coping to child adaptation (e.g. Sawyer et al., 1998; Singer et al., 2003; Thompson and Gustafson, 1996), lower maternal SOC is associated with greater patient withdrawal. Interestingly however, a cohesive family environment is also related to patient withdrawal in the present study. It is possible in this case that there is an extreme of family cohesion, where a family ‘battens down the hatches’ and itself withdraws from the world as a way of coping with their circumstances. It is possible that, while they may have a cohesive family environment, it is a maladaptive coping strategy, which has a detrimental effect on the child’s social development. Mechanic (1977) wrote of individuals who had been affected by illness or disability and their tendency to withdraw from everyday social activities and concerns. He wrote that, while such withdrawal is a natural response and can serve as a short-term coping mechanism, the continuation of withdrawal can become maladaptive. Withdrawal has a detrimental effect on social contacts and instils a sense of hopelessness, which is perhaps what is being reflected here. Some families of children affected by disability or chronic illness can have the tendency to push away the outside world and close in on themselves as a way of coping and of protecting the child, feeling that no-one else understands their predicament. This has been reported by more recent research (e.g. Speice et al., 2002; Todd, 2007) and was found to be the case with one of the families that participated in the qualitative phase of this study.

In summary then, since MPS IH affects individuals since birth, adjustment to living with the condition is an on-going process. Following BMT there is a period of semi-isolation where the child is allowed home, but contact with others is limited to minimise the risk of infection while the child's immune system is compromised. After this period, it is understandable that parents would feel overly protective of their child and would possibly avoid environments such as toddler and play groups where the potential for germs and viruses is high. However, it is possible that this lack of social integration at such a crucial time in a child's social development sets a precedent for later psychosocial adjustment, self-concept, and coping resources. Self-concept and coping resources are particularly pertinent in adolescence, as it is a time of great physical as well as psychosocial change. For adolescents affected by this MPS condition this is particularly pertinent, as progressive physical disability and short stature are for the most part more apparent at this developmental stage than at any preceding stage. Thus, visible physical differences between oneself and one's peers are an additional challenge to young people affected by this disorder, as well as changes associated with puberty. This particular group of adolescents are prone to inhibition and can be socially withdrawn, and as previous research has found with adolescents affected by orthopaedic disability, they can experience difficulties with peer relationships. As well as progressive physical disability and short stature however, it is also necessary to consider learning disability, academic failure, and adaptive competencies. The role that these factors may play in undermining the adolescent's self-esteem and self-concept, and the development of social and communication skills requires attention. Intrinsic attributes of self-esteem, self-reliance, and coping resources are particularly pertinent to the social integration and adaptive functioning of this age group, and the school experience requires further examination in relation to the psychosocial adjustment of this patient group.

Examination of patient social competency using a longitudinal design would also be very useful in identifying children that may be at risk of adjustment disorders who are affected by this condition. Relationships between early social integration and later adjustment would help with the design of appropriate intervention programmes and with the development of family and child support programmes. When providing

support to families it is important to give particular attention to the promotion of participation in group activities, particularly when the child is young and out of semi-isolation. Social integration at this time would help toward the building blocks of intrinsic coping resources. As the present and previous research has demonstrated, child psychosocial adjustment is related to parent, family, and child characteristics, as well as to aspects of a disabling condition. Thus any support initiatives aimed at optimising child quality of life and improving child psychosocial outcomes should employ a family inclusive approach. The role that parents and the family play in the MPS child's social life would also be a useful topic for future research.

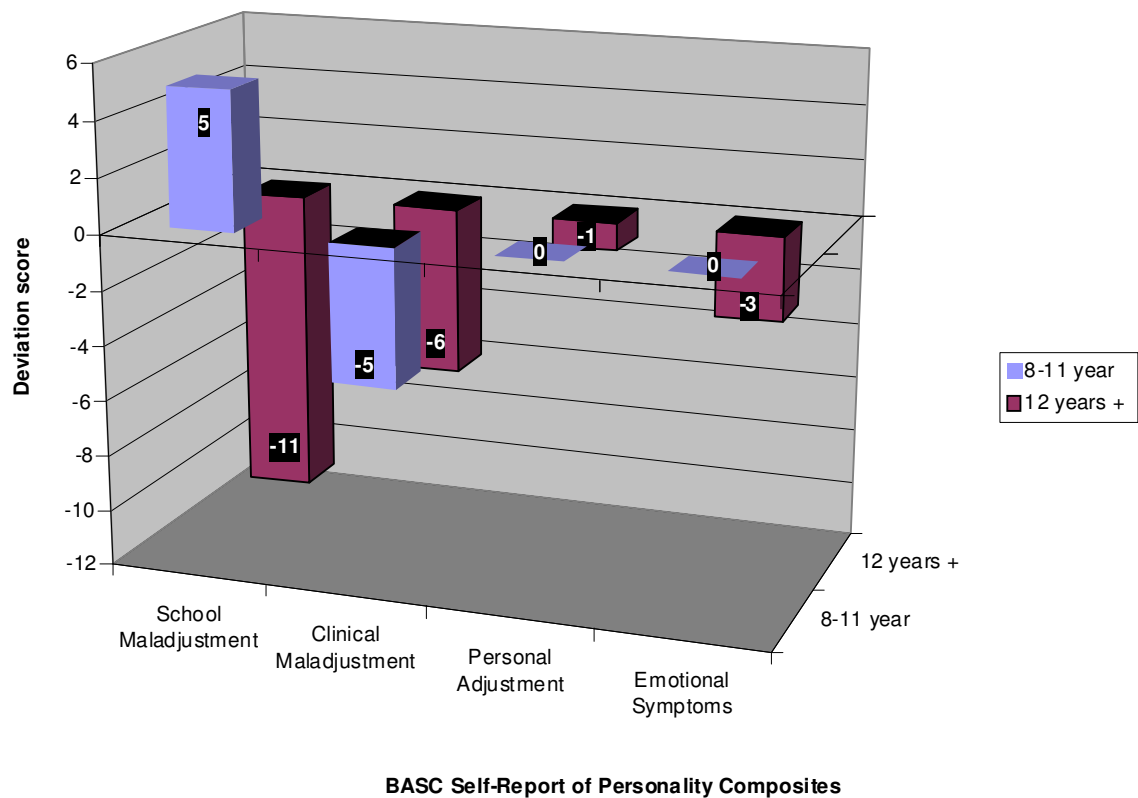
In terms of contributors to patient adjustment, a number of intrapersonal and parent stress processing and social ecological factors were found to have significant relationships with these measures of patient social competency. Furthermore, adaptive competencies were found to contribute particularly to patient social skills. Since research of this nature has not been conducted with this patient group prior to this study, it is essential that such findings are disseminated to service providers at the local level and to parents directly. This might include health care providers, particularly MPS specialist consultants, who deal with families from diagnosis throughout the child's life; and educators, who could be instrumental in encouraging autonomy, independence, a sense of achievement, and self-esteem in children and adolescents affected by this disorder. Workshops and/or written materials aimed at parents could also be useful to the support of families, particularly in the development of appropriate parenting strategies, as they adjust to having a child affected by MPS IH post-BMT.

## **7.2. Patient Self-Report of Personality: Personal Adjustment**

This section of analysis includes 18 patient participants, ten aged 8-11 years and eight aged 12 years and over, who completed the BASC Self Report of Personality (Reynolds and Kamphaus, 1998) and examines patients' scores on the Personal Adjustment composite of this measure. Nineteen patient participants did not participate because they were too young to meet the age criteria for the measure, and 7 did not participate due to the severity of their learning disability. The Personal Adjustment composite of the BASC Self Report of Personality consists of four scales: Relations with Parents, Interpersonal Relations, Self-Reliance, and Self-Esteem. This composite score is felt to be significant to the exploration of the psychosocial development of this patient population as high scores indicate positive levels of adjustment. Scores below 40 indicate possible difficulties with interpersonal relationships, self-acceptance, identity development, and ego strength. Scores below 30 are considered to be clinically significant and suggest that a child has insufficient coping skills and lacks support. Those who score low on this scale have a tendency toward withdrawal and introversion, and have difficulty with peer relations.

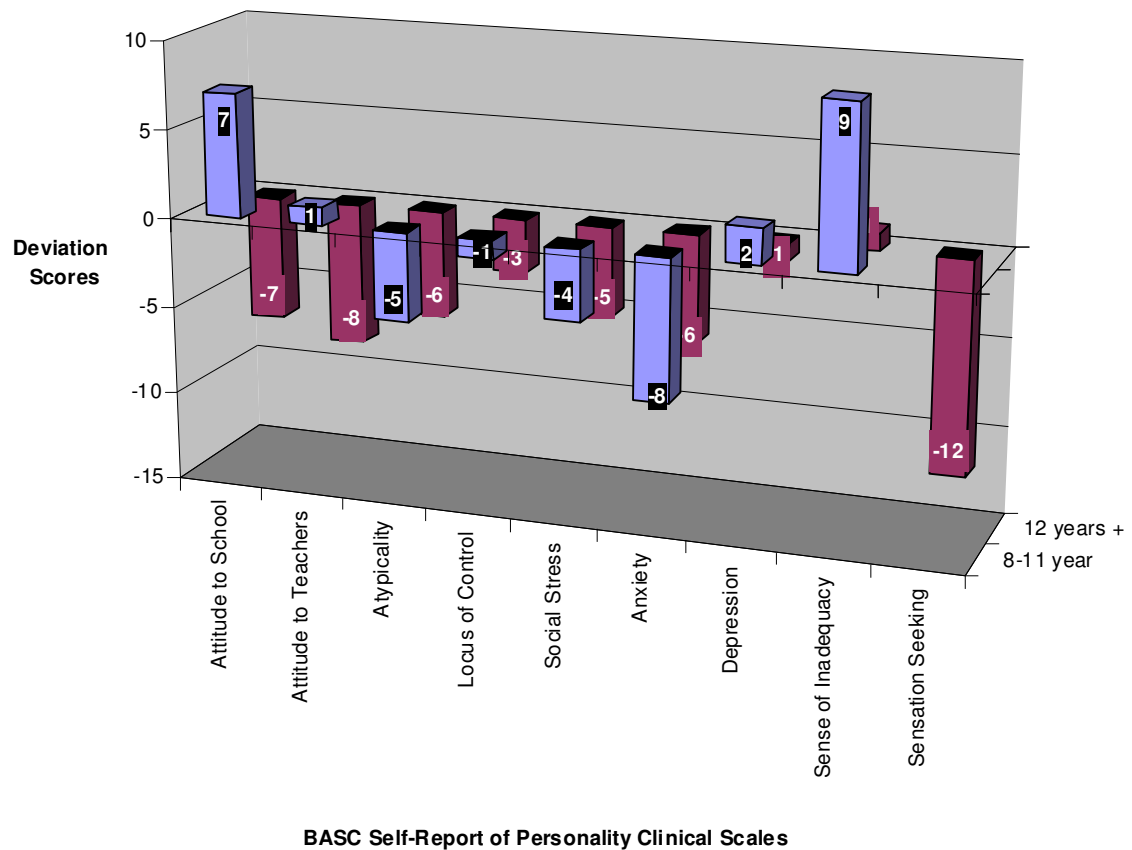
The Figures below (7-2 – 7-4) are constructed from the deviation from the mean score of the normative sample for each of the age groups (deviation score). Thus, they compare the extent to which 8-11 year olds and 12+ year olds deviate from their non-patient peers, and from each other on each of the measures. Normative scores discussed in this section refer to those set out in the BASC manual and not to the normative sample discussed in the previous section

**Figure 7-2 Comparison of the Deviation Scores for MPS Patients on Self-Report of Personality, Comparing Children aged 8-11 and Young People aged 12 years and over**



With the exception of the scores for young people aged 12+ on the School Maladjustment scale, all other scores fell within the normal range. The lower scores for this group suggest a slightly unusual acceptance of school.

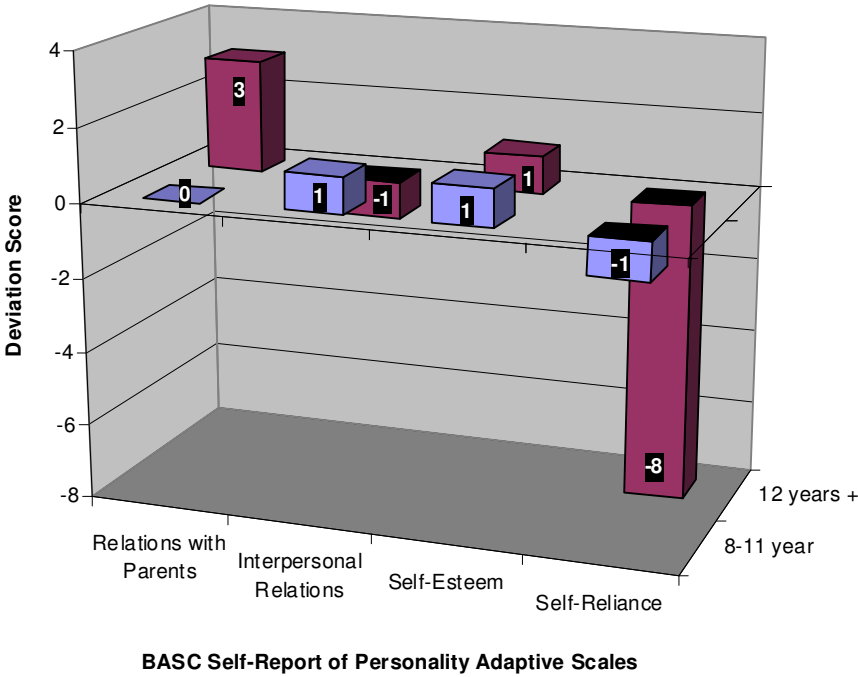
**Figure 7-3 Comparison of the Deviation Scores for the Clinical Scales between the Two Age Groups**



The comparison indicates that the mean deviation scores for the children in the 8-11 age group fall within the normal range for each scale. However, the deviation scores which approach the 'at-risk' level are Attitude to School, Sense of Inadequacy, and Anxiety, which if >10 for the first two scales and <10 for the latter scale would indicate a discomfort with school, feelings of inadequacy, particularly in academic pursuits, and the possible emergence of disruptive behaviour. Although these scores are not 'at-risk' for this patient group, they approach this level and are worthy of attention when considering educational programmes and classroom support, as they could indicate the subtle emergence of maladaptive adjustment prior to the onset of adolescence. In contrast, it is significant to note that the young people aged 12+ report considerably more favourable attitudes toward school and their teachers in comparison to both their younger peers and the normative sample. Additionally, they have a reduced sense of inadequacy in comparison to the younger age group, which

brings them more in line with the normative sample. Whilst a measure of sensation seeking was not included for the 8-11 age group, it was observed that sensation seeking is significantly lower for the 12+ age group, in comparison to a normative sample. Low scores on the Sensation Seeking scale indicate cautiousness, anxiety, inhibition, and over-controlled behaviour. It is possible that the adolescents and young adults in this patient group have a tendency toward inhibition, particularly within the school or college environment.

**Figure 7-4 Comparison of the Deviation Scores for the Adaptive Scales between the Two Age Groups**



The comparison indicated that the mean deviation scores for both children in the 8-11 age group and young people over the age of 12 years fall within the normal range. Additionally, the scores between the two age groups differ very little, with the exception of self-reliance, which appears to be slightly lower in the young people aged 12 + than both the normative sample and their younger counterparts. This could indicate a lack of self-confidence, shyness, internalisation, and insecurity. Considering this score together with those of the clinical scales, it is likely that the



children and young people in the older age group tend towards inhibition and withdrawal.

A one-way MANOVA was conducted on the four composite scores that measure School Maladjustment, Clinical Adjustment, Personal Adjustment and Emotional Symptoms Index to ascertain whether there are significant differences on these measures between the two age groups. Only one significant difference between the groups was evident, which was in relation to School Maladjustment. The older age group (mean = 39, SD = 4.02) reported significantly lower levels of school maladjustment than their younger counterparts (mean 55, SD = 13.33:  $F_{(1,16)} = 10.531$ ,  $p = .005$ ). The observed differences in age group have a large effect size, which accounts for 40% of the variance in School Maladjustment score. The means, standard deviations and eta-squared values are presented in Table 7-6 below.

**Table 7-6 MANOVA Table for the Four Composite Scores**

	Mean		SD		$\eta$
	8-11	12+	8-11	12+	
School maladjustment	54.8	38.9	13.33	4.01	.397
Clinical Adjustment	45.0	44.5	8.38	7.21	.001
Personal Adjustment	49.8	48.8	6.58	9.89	.005
Emotional Symptoms	49.8	47.5	7.33	9.56	.020

### **7.2.1. Predictive Association between Risk and Resistance Factors and Patient Personal Adjustment**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient personal adjustment than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social

ecological resistance factors. Risk factors that were added into the model were patient physical functioning and level of assistance required as measured by the MPS Health Assessment Questionnaire, and cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests. Intrapersonal resistance factors added into the model were patient adaptive skills and school maladjustment as measured by the BASC Parent Form, and socialisation as measured by the Socialisation questionnaire developed for this study. Stress processing resistance factors that were added into the model were maternal mental health as measured by the GHQ, maternal SOC as measured by the Sense of Coherence Scale, mothers' perception of child health as measured by the Child Health Questionnaire, and mothers' anxiety about child welfare as measured by the Anxiety and Risk Perceptions measure designed for this study. Social-ecological factors that were entered into the model were independent and active-recreational family environment as measured by the Family Environment Scale. The standardised regression coefficients, t-values, and probabilities for the final model is reported in Table 7-7 below to show the relative contribution of each of the child and family dimensions to patient Personal Adjustment.

**Table 7-7 Model for Patient Personal Adjustment**

Variable	Stand. Beta	t-value	Sig
<b>F = 18.741<sub>3,2</sub> p = .051 (95%)</b>			
Patient Physical Functioning	-.527	-4.122	.054
Level of Assistance Required	.622	4.778	.041*
Mother's Perception of Child's Health	-.544	-3.716	.065
Patient Cognitive Function	.522	4.476	.046*
Patient Adaptive Skills	.911	6.194	.025*
School Maladjustment	-.099	-.510	.661
Patient Socialisation	-.211	-1.575	.256
Mother's Mental Health	-.292	-1.898	.198
Maternal Sense of Coherence (SOC)	-1.373	-4.340	.049*
Mother's Anxiety about Child Welfare	-.720	-3.182	.086
Family Environment: Active-Recreational	-.935	-5.999	.027*
Family Environment: Independence Support from Friends	-.041	-.254	.823
	.413	2.946	.098

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

These relationships indicate that patient functioning, intrapersonal, stress processing and social-ecological resistance factors have the biggest impact on patient personal adjustment, accounting for 95% of the variance. The risk factor of patient cognitive function showed a significant positive relationship to patient personal adjustment, as did the level of assistance the individual required carrying out activities of daily living. The intrapersonal factor of patient adaptive skills showed a significant positive relationship to patient personal adjustment, and both the stress processing factor of maternal SOC and the social-ecological factor of family environment showed significant negative relationships to patient personal adjustment. A number of borderline significant relationships also emerged which are worthy of discussion with this small sample size. These include patient physical functioning which exhibits a negative relationship to patient personal adjustment, suggesting that patient personal adjustment is improved when physical functioning is less impaired. Two maternal stress processing factors, perceptions of child health and anxiety about child welfare, demonstrated negative relationships to patient personal adjustment, and maternal

social support, which produces a positive relationship to patient personal adjustment. These findings will be discussed.

### **7.2.2. Discussion**

The aims of this section of analysis were to explore patient self-reports of personality in comparison to a normative sample; between patients aged 8-11 and 12+ years; and to explore the contributors to patient personal adjustment. In comparison to a normative sample, patients' scores did not deviate significantly. The exception to this was the 8-11 year age group's higher scores of school maladjustment, and the 12+ year group's tendency toward inhibition, particularly in the school environment. While these findings highlight the importance of school for this sample of children, the behaviours associated with it differ between the age groups. Dissatisfaction with school, feelings of inadequacy, and disruptive behaviour are possible issues for the younger group, which may set the stage for withdrawal in adolescence if not adequately addressed at this age. Indeed, adolescents' tendency to be withdrawn and socially inhibited is illustrated here. It is possible that in adolescence, apathy in the educational environment sets in due to previous academic failure, which is a form of learned helplessness. Alternatively, or in conjunction with this, these young people may identify better with teachers than they do with peers, and thus, in their withdrawal, find school a positive experience. Secondary school is, after all, a more daunting prospect than junior school. It is bigger, has more people in it, and is more socially as well as academically demanding. Withdrawal for these young people may in fact be a form of self-preservation and a way of adapting to the additional demands being placed on them at this time.

In terms of the predictors of personal adjustment, the findings of the present study show a positive relationship between patient personal adjustment and physical and cognitive functioning, and adaptive skills. This supports previous research, which relates intrapersonal factors of social and adaptive competence (e.g. Burlew et al., 2000; Hurtig and White, 1986) and functional status, both cognitive and physical, to child psychosocial adjustment (e.g. Harper, 1983; Witt et al., 2003). The findings of the present study also highlight the relationship between patient personal adjustment and the level of assistance required by the patient carrying out activities of daily

living. More pertinently this relationship indicates that patient adjustment is more optimal the more assistance required. This relationship might be indicative of the mother's perception of the child as vulnerable as it was a subjective measure completed by the mother. It might relate to the negative relationship found between mothers' perception of child health and patient personal adjustment, which also may be indicative of maternal perceptions of child vulnerability. However, these perceptions are related to more optimal outcomes for the child in terms of adjustment, which are curious findings and do not support previous research which links maternal perceptions of child vulnerability to child adjustment difficulties. It may be possible that these relationships show the positive consequences of the mother-child caring relationship, suggesting that nurturing attention that is related to the child's health condition results in a feeling of well-being in the child. This has implications for the individual's identity formation in terms of the 'sick role' (Parsons, 1951) and requires further exploration.

In terms of parent stress processing factors, the present study shows a negative relationship between maternal SOC and patient personal adjustment, which suggests an association between positive child outcome and poor maternal coping resources. This is an unexpected finding, as previous research has related low maternal assertiveness, self-efficacy, and helplessness to poorer child psychosocial outcomes (Thompson et al 2002). It is possible that maternal SOC is low due to the uncertain nature of the long-term outcomes of BMT for this condition. Mothers may find it difficult to see their children's lives as predictable, and this lack of predictability may render the perception of the situation less manageable. It could be argued that the inability to see too far into the future puts the emphasis on today and on the immediate needs, welfare, and happiness of the child. Indeed, this potentially supports previous research, which highlights parents' tendency to live 'one day at a time' when they have children with disabilities (Ray, 2002); and which illustrates mothers' tendency to prioritise their role as mother in their child's adaptation (Pelchat et al 2003). Living each day as if it were the last may have a positive effect on the child's adjustment, although the caveat here is that failure to parent with a future in mind can have implications for an individual's ability to socially mature (McAnarney, 1985; Orr et al 1984).

Another interesting and unexpected finding is the indication of a negative relationship between the social-ecological factor of family environment that is active-recreational orientated and patient personal adjustment. This does not support previous research which has linked active-recreational orientation with healthy psychosocial adjustment in children with disabilities (e.g. Moos & Moos, 2002, Nihira et al, 1980, Nihira et al, 1981). One possible explanation for this anomalous result lies with the items on the Active Recreational Orientation scale of the Family Environment Scale (Moos & Moos, 2002), which ask whether *overall* or *for the most part* family members enjoy hobbies, sports, and going out. It may be possible that for the most part family members do not enjoy such recreational activities but that the child affected by MPS IH does, and that the emphasis is put on the child's enjoyment of activities rather than on other members of the family. Thus, the child's active recreational activities were not effectively recorded using this measure. On the other hand, it may be that family members on the whole do enjoy recreational activities, but that the child affected by MPS IH is not afforded the enjoyment of activities that are to his or her personal taste. It is possible that parents may include the child in a variety of recreational activities that they or other siblings enjoy, but which the child affected by MPS IH does not. Moreover, it is possible that child psychosocial adjustment is optimised by families that facilitate the enjoyment of the child's interests rather than including the child in activities enjoyed by the family majority.

In summary then, patient personal adjustment when the individual is affected by MPS IH post-BMT appears to be related to a supportive family environment where the child's health and care needs are adequately catered for, and where adaptive and social skills are allowed to develop through the development of autonomous behaviour, feelings of adequacy, and social and recreational experiences that are chosen by, and of interest to, the child. It is also related to parenting that meets the physical and emotional needs of the child without being over protective and fostering dependence. Organisations that are involved in providing support to such families need to be aware of such findings so that appropriate support can be given to families as early as possible, particularly in terms of parenting. Although living 'one day at a time' may have some benefits for the development of confidence and self-esteem, it is also important to consider the future adult needs of the individual affected by this disorder, as adjustment disorders, particularly in terms of withdrawal and inhibition

are apparent for this patient group. More candid information from medical professionals regarding longevity post-BMT would benefit parents enormously by reducing their perception that their child continues to be vulnerable in terms of their mortality. The results also show how the school environment impacts on the child, and how different issues may be pertinent to children as they develop into adolescence and move from primary to secondary school. In the 8-11 year age group, awareness of differences between oneself and one's peers becomes apparent, and learning difficulties as well as physical limitations are relevant for this patient group. Appropriate and consistent classroom support is essential for this group of children so that a sense of adequacy, achievement, and self-esteem can be achieved. In terms of adolescence, this is a difficult time for these young people, as physical disability progresses. Special attention needs to be given to young people as they enter secondary school and to appropriate classroom support and social activities, and the potential consequences this has for individuals psychosocially and educationally require attention. Overall, to afford a child affected by MPS IH the opportunity to adjust well post-BMT attention needs to be given to these issues within the home as well as within the school environment; and psychosocial support within the school environment should be considered.

### 7.3. Patient Cognitive and Adaptive Function, and Education

#### 7.3.1. Cognitive and Adaptive Function

Cognitive function varies enormously for this patient group. In order to accommodate the wide range of function and the wide age range of the population, three tests of cognitive function were used. Each scale has age parameters, which were adhered to unless the scale was not appropriate for the patient's abilities. The normative mean for the Griffiths is 100.18 and the standard deviation 12.76. Thus, as can be seen in Table 7-8 below, the children's abilities ranged from profound deficit to within the normal range. Similarly for the WISC-III and the WAIS-III, which have normative means of 100 and standard deviations of 15, the scores for the MPS IH post-BMT sample ranged from significant delay to within the normal range, as can be seen in Table 7-8 below.

**Table 7-8 Descriptive Statistics for Griffiths Mental Development Scales**

Scale	N	Min	Max	Mean	Std. Dev
Griffiths GQ Standard Score	23	5.48	98.00	65.30	25.88
Locomotor sub-quotient	23	7.10	99.00	55.69	25.67
Personal-Social sub-quotient	23	8.10	114.28	70.99	26.29
Hearing & Speech sub-quotient	23	6.20	131.43	69.02	33.96
Eye & Hand Coordination sub-quotient	23	3.70	90.00	64.00	24.11
Performance sub-quotient	23	2.30	101.70	64.90	24.86
Practical Reasoning sub-quotient	13	23.00	118.00	68.69	32.29

The Locomotor subscale of the Griffiths Mental Development Scales measures gross motor skills including the ability to balance, co-ordinate and control movements. The Personal-Social subscale measures the child's proficiency in activities of daily living, their level of independence and interaction with other children. The Hearing and Speech subscale assesses receptive and expressive language. The Eye and Hand Co-



ordination subscale measures fine motor skills, manual dexterity and visual monitoring skills. The Performance scale assesses visual-spatial skills including speed of working and precision, and the Practical Reasoning subscale assesses the child's ability to solve practical problems, and their understanding of basic mathematical concepts and moral issues. The first five scales assess development from infancy to age eight years, and the last scale from three to eight years.

Table 7-8 above illustrates how the patient participants whose ability was assessed by the researcher using the Griffiths Mental Development Scales scored particularly poorly on the locomotor subscale in comparison to the other subscales, in particular the Personal-Social and Hearing and Speech subscales, indicating significant delay in this area. This is not particularly surprising considering the nature of the MPS condition and its progressive bone and joint disease, and the implications this has for orthopaedic impairments and physical functioning. The data also illustrate how patient participants showed significant developmental delay as the mean total GQ score, the total score for the Griffiths scales as a whole, is almost three standard deviations below the standardised mean. This is also the case for all of the sub-quotients. A one-way repeated measures ANOVA was conducted to compare mean scores on the Griffiths sub-quotients with the exception of the Practical Reasoning sub-quotient as it reduced the number of participants in the analysis to 13 and would compromise the power of the test. There was a significant effect for subscale (Wilks' Lambda = .47,  $F(4, 19) = 5.28$ ,  $p = .005$ , multivariate eta squared = .53).

Pearson product-moment correlations revealed a number of significant positive relationships between the Griffiths sub-quotients and patient and parent adjustment outcomes. Internalising problems for patient participants aged 2-5 years as measured by the BASC parent form were found to be positively related to the Griffiths Personal-Social ( $r = .771$ ,  $n = 12$ ,  $p = .003$ ), Hearing and Speech ( $r = .623$ ,  $n = 12$ ,  $p = .023$ ), Hand-Eye Coordination ( $r = .592$ ,  $n = 12$ ,  $p = .042$ ), Performance ( $r = .795$ ,  $n = 12$ ,  $p = .002$ ), and Practical Reasoning ( $r = .763$ ,  $n = 7$ ,  $p = .046$ ) sub-quotients. This suggests that the better the development in these domains the greater the child's internalising problems.

In terms of parent participant outcomes significant relationships were found between maternal mental distress as measured by the Child Abuse Potential Inventory (Milner,

1986) and the Griffiths Personal-Social sub-quotient ( $r = .439$ ,  $n = 23$ ,  $p = .036$ ). Maternal risk perceptions when they had children aged 5 years and under were found to be negatively related to the Griffiths Performance IQ ( $r = -.485$ ,  $n = 17$ ,  $p = .048$ ). Maternal expectations of their child's future achievements were also positively related to the Griffiths Personal-Social ( $r = .458$ ,  $n = 22$ ,  $p = .032$ ), Hearing and Speech ( $r = .610$ ,  $n = 22$ ,  $p = .003$ ), Hand-Eye Coordination ( $r = .486$ ,  $n = 22$ ,  $p = .022$ ), Performance ( $r = .455$ ,  $n = 22$ ,  $p = .033$ ), and Practical Reasoning ( $r = .644$ ,  $n = 13$ ,  $p = .018$ ) sub-quotients. These relationships indicate that mothers experience more mental distress the higher their child scores on the Personal-Social subtest items, they perceive their child to be less at risk of coming to harm when carrying out everyday childhood activities the higher they score on the Performance subtest items, and have higher expectations of the child the higher they score on all subtests of the measure.

**Table 7-9 Descriptive Statistics for WISC-III and WAIS III**

Scale	N	Min	Max	Mean	Std Dev
<b>WISC and WAIS</b>					
Full Scale IQ	21	40.00	114.00	67.38	18.08
Verbal IQ	21	46.00	128.00	74.67	19.12
Performance IQ	21	46.00	104.00	65.86	15.62
<b>WISC</b>					
Verbal Comprehension IQ	16	50.00	127.00	76.81	20.43
Perceptual Organisation IQ	16	50.00	100.00	66.12	15.03
Freedom from Distractibility	16	50.00	109.00	73.87	17.69
Processing Speed	13	50.00	117.00	69.61	17.20
<b>WAIS</b>					
Verbal Comprehension Index	5	74.00	96.00	80.00	9.27
Perceptual Organisation Index	5	60.00	118.00	80.40	22.24

Verbal IQ on the WISC and WAIS is calculated from scores achieved on verbal subtests which consist of oral questions. These include giving oral answers to general information questions, explaining how two things or concepts could be alike, orally solving arithmetic problems, and giving oral definitions of words. Performance IQ is calculated from scores achieved on performance subtests which consist of non-verbal problems. These include identifying the missing parts of a picture, making rows of shapes with different lines according to a code, sequencing cartoon pictures to make sensible stories, copying geometric designs with plastic cubes, searching for a target symbol in a row of symbols, and the completion of puzzles and mazes. Full Scale IQ

is based on the ten tests included in the Verbal and Performance (nonverbal) IQ scales.

Overall, patient participants who completed these scales appear to demonstrate cognitive functional impairment as the mean scores on all scales and subscales fall between one and three standard deviations below the normative mean. Table 7-9 however illustrates how the patient participants who completed the WISC scored higher on the subscales that comprised verbal tasks, including Verbal Comprehension, and lower on the subscales that comprised non-verbal tasks, for example Processing Speed. Those who completed the WAIS on the other hand scored equally well on the Verbal Comprehension and Perceptual Organisation Indexes. A paired samples t-test was conducted between patient participants' mean scores of Verbal IQ and their mean scores of Performance IQ and a significant difference was found, with participants achieving higher scores on Verbal IQ ( $M = 74.67$ ,  $SD = 19.11$ ) than on Performance IQ ( $M = 65.86$ ,  $SD = 15.62$ ,  $t(20) = 3.33$ ,  $p < .005$ ). Again, this is not a surprising result as individuals affected by MPS IH post-BMT have poor eyesight and have difficulty manipulating objects with their hands due to joint restrictions in their fingers. This difficulty increases with age as bone and joint disease progresses.

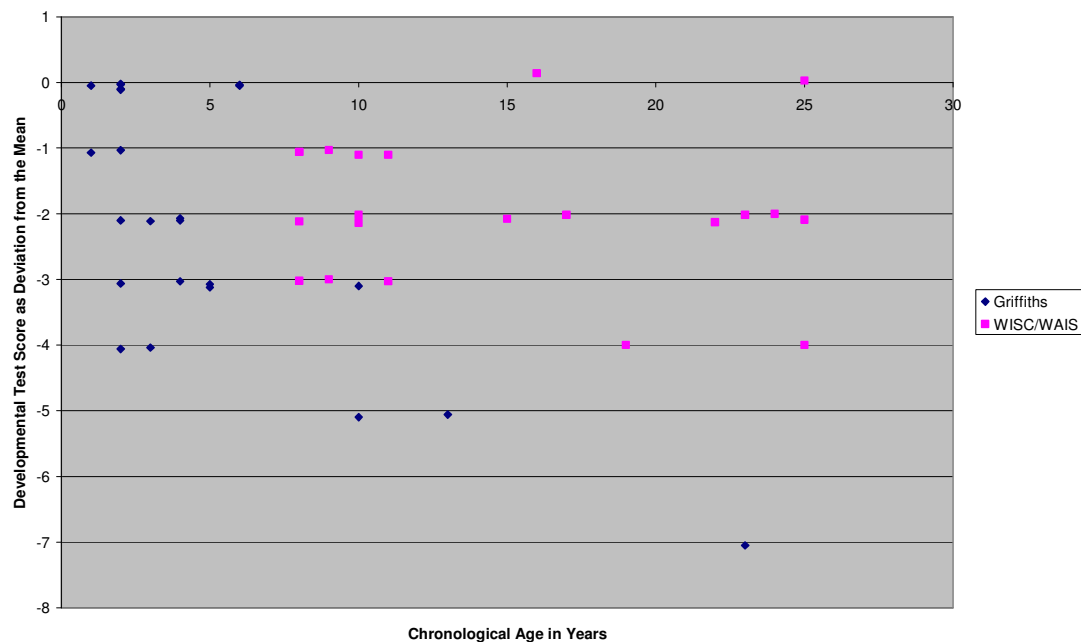
Pearson product-moment correlations were also computed and a number of significant relationships were noted. Verbal IQ ( $r = .535$ ,  $n = 21$ ,  $p = .013$ ), Performance IQ ( $r = .455$ ,  $n = 21$ ,  $p = .038$ ), and Full Scale IQ ( $r = .545$ ,  $n = 21$ ,  $p = .011$ ) as measured by the WISC and WAIS showed significant positive relationships to mothers' perception of child health. Maternal perception of the impact the child's condition had on the family also showed a significant positive relationship to Verbal IQ ( $r = .465$ ,  $n = 21$ ,  $p = .034$ ), Performance IQ ( $r = .493$ ,  $n = 21$ ,  $p = .023$ ) and Full Scale IQ ( $r = .467$ ,  $n = 21$ ,  $p = .033$ ) as measured by the WISC and WAIS. These relationships indicate that mothers have more positive perceptions of their child's health and perceive their child's condition to impact less on the family when the child's IQ is higher.

In terms of parent stress and coping maternal Sense of Coherence was found to be positively related to Full Scale IQ ( $r = .459$ ,  $n = 20$ ,  $p = .042$ ), and SOC meaningfulness to Verbal IQ ( $r = .481$ ,  $n = 20$ ,  $p = .032$ ), Performance IQ ( $r = .457$ ,  $n = 20$ ,  $p = .043$ ) and Full Scale IQ ( $r = .524$ ,  $n = 20$ ,  $p = .018$ ) as measured by the

WISC and WAIS. Parent distress was found to be negatively related to Performance IQ ( $r = -.492$ ,  $n = 20$ ,  $p = .027$ ) and Total IQ ( $r = -.490$ ,  $n = 20$ ,  $p = .028$ ) as measured by the WISC and WAIS. These relationships indicate that maternal SOC in general is higher when their child's overall IQ is higher. They also indicate that mothers' ability to perceive their world as meaningful and demands as challenges, worthy of investment and engagement are related to child IQ on all sub-scales of the WISC and WAIS; and that maternal distress that is directly related to parenting and the parent-child system is lower when the child's Performance IQ is higher.

Figure 7-5 shows the distribution of participants' total scores from all three of the developmental scales. For this purpose, cognitive function has been calculated as standard deviations from the mean, so that all three measures of cognitive function could be included in the same variable, and thus all participants. The majority of original scores for both the WISC and the WAIS were between 50 and 80, with 4 outliers, 2 at 40 and 2 above 100. Original total scores on the Griffiths test formed two clusters, one between 80 and 100, and another between 40 and 70. Only three individuals scored below 30, with one scoring particularly low, and these were three of the older participants who completed this scale.

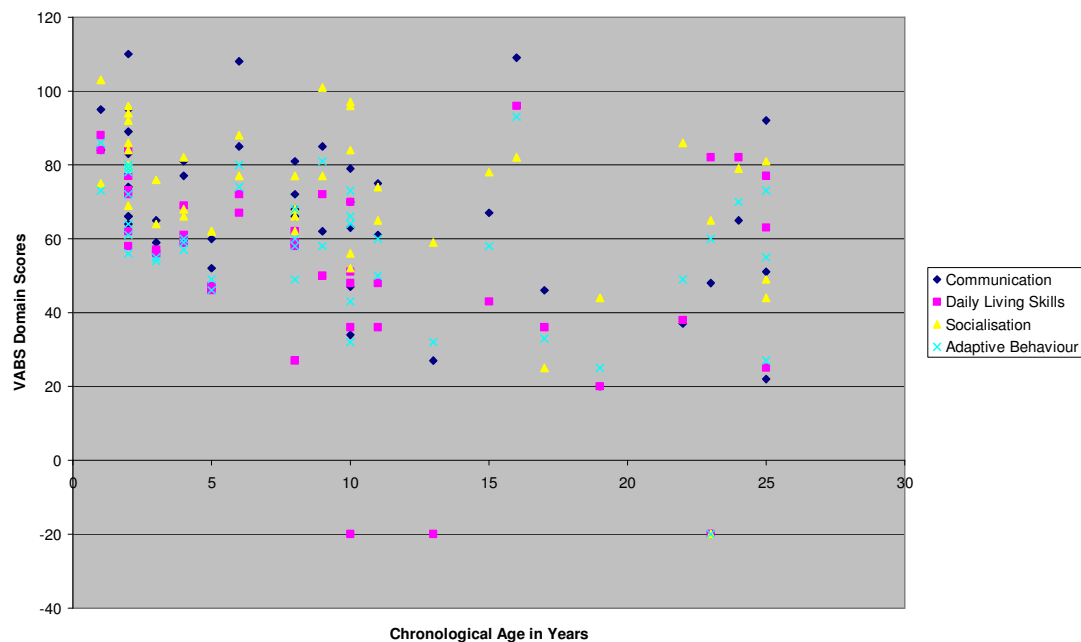
**Figure 7-5 Distribution of Total Scores for Griffiths Mental Development Scales, WISC-III, and WAIS-III as Standard Deviations from the Scales' Normative Means**



The findings of the present study reveal a significant negative correlation between the patient's age at transplant and cognitive function (Pearson's  $r = -.320$ ,  $p = .036$ ), when cognitive function is calculated as standard deviations from the normative mean. They also show a significant negative relationship between adaptive function and age at transplant (Pearson's  $r = -.383$ ,  $p = .011$ ). Patient age at transplant was calculated in months and was limited to the last BMT the child underwent or the BMT that successfully engrafted, be that the first or second transplant.

When looking at the distribution of cognitive function scores in terms of patient age, they appear to be more spread out amongst the older patients. This also seems to be the case for the adaptive function measures on all of the Vineland Adaptive Behaviour Scales (VABS) (Sparrow et al., 1984) domains, indicating a wider range of ability amongst the older patients. This is illustrated in Figure 7-6.

**Figure 7-6 Distribution of Scores for the Vineland Adaptive Behaviour Scales' Communication, Daily Living Skills, Socialisation and Adaptive Behaviour Domains**



In order to further explore the apparent differences between the older and younger patients, the group was split at the age of 12. The rationale for this choice is that the population is naturally more heavily loaded in the under-12 age group, and those aged over 12 are for the most part nearer to adulthood than childhood, as illustrated in Table 7-10.

**Table 7-10 Descriptive Statistics for the Proposed Age Split**

Age Group	N	Min Age	Max Age	Mean Age	Modal Age	St Dev
< 12 years	32	1	11	5.62	2	3.45
12 years +	12	13	25	20.58	25	4.36

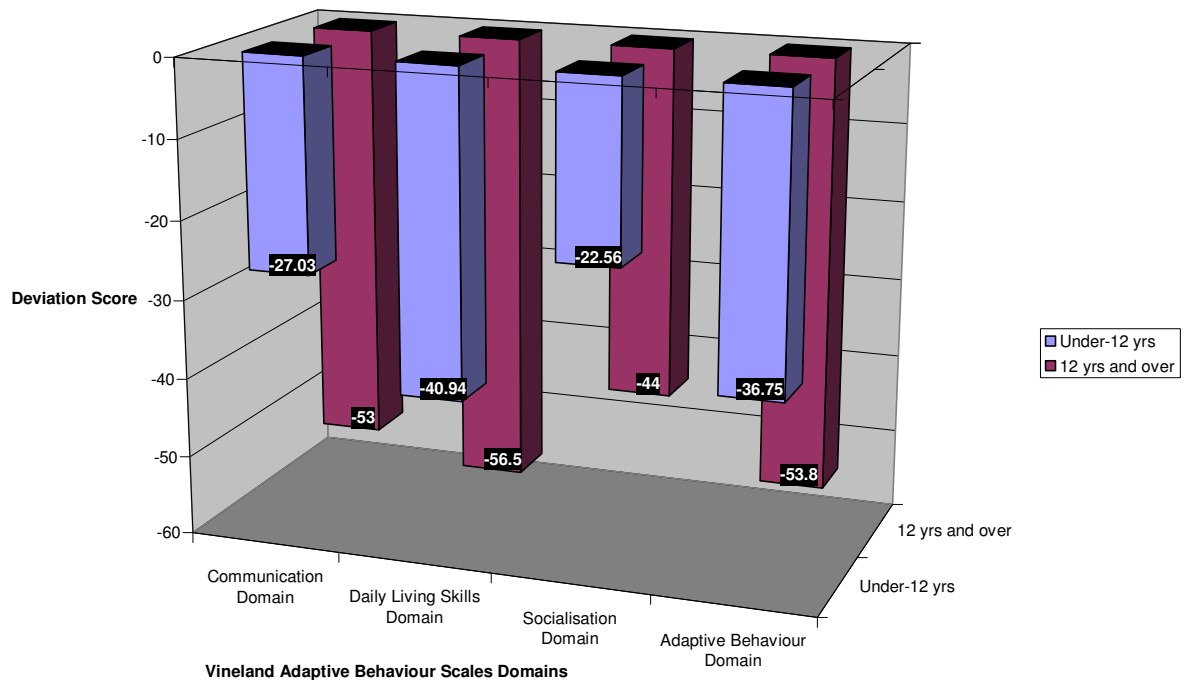
Using an independent samples t-test, no differences were found between the mean scores of cognitive function for the two age groups. A one-way MANOVA was conducted on three of the four composite scores of the VABS however, and on the overall Adaptive Behaviour score, which revealed significant differences between the two age groups in relation to Socialisation, Communication, and overall Adaptive

Behaviour. The older age group reported significantly lower levels of Socialisation ( $M = 56$ ,  $SD = 30.71$ ), Communication ( $M = 47$ ,  $SD = 34.32$ ), and Adaptive Behaviour ( $M = 46.25$ ,  $SD = 29.33$ ) than their younger counterparts (Socialisation:  $M = 77.44$ ,  $SD = 13.63$ :  $F = 10.441_{(1,42)}$ ,  $p = .002$ ; Communication:  $M = 72.97$ ,  $SD = 16.46$ :  $F = 11.574_{(1,42)}$ ,  $p = .001$ ; Adaptive Behaviour:  $M = 63.25$ ,  $SD = 12.87$ :  $F = 7.251_{(1,42)}$ ,  $p = .010$ ). The observed differences in age group have moderate to large effect sizes, which account for 20% of the variance in Socialisation, 22% in Communication, and 15% in Adaptive Behaviour. The means, standard deviations and eta-squared values are presented in Table 7-11 below. Figure 7-7 also illustrates how the patients in the younger age group demonstrate better adaptive function on each of the VABS domains explored, than their older counterparts. It also illustrates however, that this patient group as a whole has lower functioning in terms of adaptive skills than a normative population.

**Table 7-11 MANOVA Table Comparing VABS Domain Means for the two Age Groups**

	Mean		SD		$\eta^2$
	<12s	12+	<12s	12+	
Communication	72.97	47.00	16.46	34.32	.216
Daily Living Skills	59.06	43.5	20.8	38.46	.066
Socialisation	77.44	56.00	13.63	30.71	.199
Adaptive Behaviour	63.25	46.25	12.87	29.33	.147

**Figure 7-7 Comparison of the Deviation Scores for the VABS Domains between the Age Groups**



### 7.3.2. Predictive Association between Risk and Resistance Factors and Patient Cognitive Function

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient cognitive function than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) Illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. The choice of variables was informed by previous research into BMT outcomes for MPS patients, modelling-theoretical assumptions in relation to the chronic illness literature, and from statistical calculations from correlational analyses. Risk factors that were added into the model were patient age at last BMT, complications experienced by the child during and post-BMT as rated by the mother in the parent interview, how stressful the BMT was rated by the mother in the BMT Perceptions measure developed for this study, the number of telephone calls or visits made to the GP or MPS consultant in the last 6 months, and the number of



calls or visits made to the specialist MPS hospital in the last 12 months as measured by the Medical History Questionnaire, and the level of assistance required by the patient carrying out activities of daily living as measured by the MPS I Health Assessment Questionnaire. The intrapersonal factor of patient adaptive skills as measured by the BASC Parent Form was added into the model. Stress processing resistance factors that were added into the model were mothers' perception of child health as measured by the Child Health Questionnaire, maternal spiritual beliefs as measured by the Beliefs and Attitudes Scales developed for this study, maternal anxiety about child welfare as measured by the Maternal Anxiety and Risk Perceptions Questionnaire developed for this study, and mothers' expectations of child as measured by the Expectations Questionnaire also developed for this study. Two social-ecological factors were added into the model, which pertained to family utilitarian resources. These were mothers' education and job classification as reported by mothers in the parent interview. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 7-12 below to show the relative contribution of each of the BMT, child, and parent dimensions to patient cognitive function.

**Table 7-12 Model for Patient Cognitive Function**

Variable	Stand. Beta	t-value	Sig
<b>F = 5.733<sub>3,22</sub>, p = .005 (65%)</b>			
Child's Age at Last BMT	.143	1.170	.255
BMT Complications	.025	.159	.875
BMT Stress	-.003	-.019	.985
MED 1: Calls/Visits to GP	.255	1.967	.062
MED 2: Calls/Visits to MPS Hospital	.377	2.205	.038*
Mother's Perception of Child's Health	.161	1.092	.287
Patient Adaptive Skills	.004	.031	.976
Level of Assistance Required	-.570	-3.417	.002**
Mother's Education	.009	.067	.947
Mother's Job Classification	-.477	-3.687	.001**
Maternal Beliefs	-.213	-1.674	.108
Maternal Anxiety about Child Welfare	.076	.477	.638
Mother's Expectations of Child	.470	3.480	.002**

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

With all variables entered into the analysis, 65% of the variance of patient cognitive function was explained by patient functioning and illness-related stressors, and stress processing and social-ecological resistance factors. The risk factor of the level of assistance required by the patient was negatively related to cognitive function, and the number of visits to the MPS hospital showed a significant positive relationship to patient cognitive function. The number of telephone calls and visits to the GP and MPS consultant in the last 6 months also showed a positive relationship to patient cognitive function though this was of borderline significance. The stress processing factor that showed a relationship to patient cognitive function was mothers' expectations of the child, which showed a significant positive relationship. Similarly, the social-ecological factor of mothers' job showed a significant negative relationship to patient cognitive function, indicating that the higher the job classification the better the child's cognitive functioning. These findings thus indicate relationships between patient cognitive function and: i) The amount of medical attention the child receives; ii) Child self-reliance in terms of activities of daily living; and iii) aspects of the mother in terms of her expectations of the child and her past or present job classification. These relationships will be discussed in relation to past and future research, and to practical applications.

### **7.3.3. Education**

With regard to education, and the patient participants of the present study, eight young adults were no longer in full time education. However, one went to mainstream school with no educational support at primary or secondary level; two attended a special needs unit within a mainstream school for both primary and secondary school; four went to special needs schools throughout their education, and one was in mainstream education until the second year of secondary school, and special needs education thereafter. Thus, 50% of the young adults in this population went to special needs school, 37.5% went to mainstream school, and 12.5% had both mainstream and special needs education. Five children and young people were being educated at secondary school level: One was in mainstream school receiving assistance for physical disability only; two were in mainstream school with one-to-one learning support; and two were in special needs schools. Forty percent of the young people currently at secondary school therefore went to special needs school. Seventeen

children were in primary school. Ninety-four percent of them attended mainstream school with educational support, and 6% went to special needs schools. Six children were in nursery education. Sixty-seven percent attended mainstream nursery with support, while 33% attended special needs nurseries.

Mothers were asked to rate from 1 to 7 how satisfied they were with their child's schooling from three different perspectives: Academically, socially, and emotionally. A rating of 1 on the scale denoted complete dissatisfaction, while a rating of 7 denoted complete satisfaction. Overall parents were quite satisfied with their children's education from all three perspectives, and there were no significant differences between each of the scale dimensions when explored using an unrelated one-way analysis of variance. Thus, mothers were no more or less satisfied with one aspect of their child's schooling compared to another. Academically 77.8% of mothers rated their satisfaction over the half-way point on the 1-7 scale; Socially 80.6% of mothers rated their satisfaction over the half-way point on the 1-7 scale; and Emotionally 69.5% of mothers rated their satisfaction over the half-way point on the 1-7 scale. Descriptive statistics for each of the scale dimensions and total scale scores are shown in Table 7-13 below.

**Table 7-13 Descriptive Statistics for Satisfaction with Schooling Scale**

Satisfaction with Schooling	N	Min	Max	Mean	Std. Dev
Academic	36	1.00	8.00	5.42	1.56
Social	36	1.00	7.00	5.50	1.76
Emotional	36	1.00	8.00	5.64	1.81
TOTAL	36	6.00	21.00	16.67	4.26

#### **7.3.4. Discussion**

The findings of the present study show the younger patients to have better adaptive function than their older counterparts in terms of socialisation, communication and overall adaptive function. Although the difference between the age groups was not found to be statistically significant, there is also an indication that cognitive function

may be slightly better amongst the younger patients. The reasons for this apparent improvement in function are not clear, as there are so many possible contributors to this type of functioning. These include pre-transplant function, GvHD, infections, and other medical complications. It is also likely that the percentage of engraftment achieved, the type of transplant given, and factors associated with the donor have an impact on cognitive and adaptive functional outcome. Furthermore, BMT regimens have changed dramatically since it was an experimental treatment for MPS IH in the late 1970s, which could contribute to an overall improvement in cognitive and adaptive functional outcome. However, since all of the information gathered for this piece of research was gleaned from parents, much of this additional information was inaccurate or not known, and was therefore not deemed reliable enough or sufficient for inclusion in this analysis.

However, what did emerge from the data was the relationship between cognitive and adaptive function and age at engrafted BMT, which further advocates the dissemination of information about rare genetic disorders to front-line medical professionals, so that appropriate referral, diagnosis and treatment can be achieved as early as possible in the child's life. It also supports medical research which has found developmental trajectories to be more favourable when the child is transplanted at a younger age, preferably under 12 months (e.g. Hopwood et al., 1993; Shapiro et al., 1995; Peters et al., 1998; Escolar and Kurtzberg, 2002). It should be noted however, that for the present patient sample, the range of both cognitive and adaptive function remained to be large for those transplanted under-12 months and for those transplanted between 12 and 24 months. This suggests the involvement of factors other than just age at transplant in cognitive functional outcome.

Much research into cognitive and neurological function post-BMT for MPS IH patients has recorded the stabilisation of IQ and improvements post-BMT (e.g. Peters et al., 1996; Shapiro et al., 1995) and cord blood transplants (Staba et al., 2004). However, in two cases of adolescents affected by this disorder, a slight decline in cognitive function has been recorded as the individuals got older (Hopwood, et al., 1993). It is not clear if this is an age effect however, or whether other psychosocial factors are involved, which render the individual withdrawn, particularly in an educational environment. As discussed in the literature review, a significant

percentage of children and adolescents affected by chronic illness experience problems at school, which are of a psychosocial nature (Wray et al., 2001); and social and behavioural problems have been found to mask intellectual ability (Whittington et al., 2004). It is possible then that inhibition and withdrawal in adolescence is mistaken for learning inadequacy in some instances in this patient group. Academic failure in children with learning difficulties can also lead to further academic failure, as an individual's self confidence and motivation to continue to learn is depleted (Hersh et al., 1996). This form of 'learned helplessness' could appear as a loss of IQ as the child moves into adolescence and the more challenging environment of secondary school. As previously discussed and noted in the findings of the present study, this may be more pertinent for children with more moderate learning difficulties as they may be aware of their limitations in comparison to their peers. Psychosocial functioning in adolescence may also be reflective of such functioning in younger childhood. Indeed, this is supported by the correlational analyses conducted which highlighted positive relationships between patient internalising problems and all sub-quotients on the Griffiths Mental Development Scales. Psychosocial issues therefore require attention in the school environment, so that opportunities for continued educational progress are maintained into adolescence and early adulthood.

On examination of the subtests and sub-quotients of the developmental tests utilised, it was revealed how patient participants performed significantly less well on the items and subtests that required physical activity and the physical manipulation of objects or the use of a pen/pencil. The majority of these tasks were timed and the opportunity to gain extra points, particularly on the WISC and WAIS, is given for speedy responses. Individuals affected by MPS IH post-BMT are significantly hindered in the completion of such tasks as they have orthopaedic difficulties and can have poor eyesight and stiff finger and wrist joints, which make physical activity, the manipulation of objects, and holding a pen difficult. Amongst the patient participants who completed the Griffiths Mental Development Scales, although mean scores on the Personal-Social and Hearing and Speech sub-quotients were significantly below the standardised mean, the highest scores achieved exceeded the standardised mean by over one to three standard deviations. Five children achieved a score of 100 or over on these subscales and they were aged 1 and 2 years old on the Personal-Social sub-quotient, and from 1 to 6 years on the Hearing and Speech sub-quotient. They were

therefore not necessarily the same children. For some of the Personal-Social items for very young children parents are asked to give responses regarding their child's social competence and abilities in other personal and social areas. These scores could therefore have shown a parent response bias.

The multiple regression analysis revealed factors other than those related to the child's medical history to demonstrate a relationship with cognitive function post-BMT. Interestingly, the better the child's cognitive function, the more phone calls and visits were made to the GP, and the more routine visits were made to the MPS specialist hospital. It may be possible that more visits are made to the GP as a result of the child being better able to articulate feelings and express him or herself when feeling unwell or in pain. Routine visits to the specialist hospital, if more frequent than for regular checks, could indicate the ongoing monitoring of a situation such as the changing angles of joints with a view to orthopaedic surgery. A child with better cognitive function may feel more strongly about having corrective surgery done, so that they can continue to be mobile and participate in physical activities. Alternatively, parents may consider a child's quality of life and self-concept more in relation to physical functioning when they have better cognitive function, and may put more emphasis on maintaining physical functioning so that the child continues to be socially active. With better cognitive function more emphasis may also be put on self-reliance and autonomy. Thus, physical functioning is perhaps more of a priority for families with children with better cognitive function, so that independence is prolonged.

Continuing with the topic of patient self-reliance and autonomy, the results also show a negative relationship between patient cognitive function and the level of assistance required carrying out activities of daily living. While this result may simply illustrate a relationship between higher cognitive function and better-developed adaptive skills, it may also have implications for the promotion of resourcefulness and self-help activities, which have been related to improved quality of life in individuals affected by chronic illness and disability (Braden, 1990; Rosenbaum, 1983). This finding has implications for parenting, and potentially supports previous research, which has related a non-assertive, passive, and depressive parenting style to a decline in child cognitive function over time (Thompson et al., 2002). Child cognitive function is also

linked to mothers' job classification here, a social-ecological resistance factor, which relates to family utilitarian resources. This is an interesting finding as child IQ has previously been linked to maternal IQ or level of education achieved rather than to her job (Armor, 2003). However, maternal resourcefulness, professional ability, organisation, and leadership qualities may be more relevant to child development on a day to day level in terms of this condition than to past academic achievements.

Indeed, previous research has found characteristics of the family such as income and mother's education and age to be related to maternal stress (Wallander et al., 1998), and maternal knowledge about child development (Thompson et al., 2002) and cognitive growth fostering (Fulton et al., 2003) to be related to lower levels of maternal distress. In turn utilitarian family resources have been related to child psychosocial outcomes (Wallander et al., 1989). It is suggested that more highly educated mothers are better able to access social resources and information, have more knowledge of resources, and to be more organised. Such resources are purported to enable mothers to more adequately manage 'tangible everyday nuisances' before they impact on them psychologically (Manuel, 2001). Furthermore, a mother who has perhaps given up a good career to care for her child may be more proactive in terms of helping the child to progress. She may be more inclined to use problem-solving skills than a mother that has not had a serious career; and she may be more likely to spend time researching the disorder and outcomes of BMT, as well as health management and education programmes. She may also be more able to provide the child with a stimulating environment. These issues may also be relevant to the physical management of the condition, and may partly explain why a child with better cognitive function may seek more medical attention. In this case the relationship may be more pertinent to characteristics of the mother than to those of the child, and may be indicative of her proactive management strategies in terms of the child's health and/or physical functioning status.

Finally, patient cognitive function was related to the stress processing factor of mothers' expectations of child future achievements. While it is logical that mothers of children with better cognitive function would have higher expectations of their child, mothers of children with disabilities have been found to have low expectations of their children, particularly in terms of future independence from the family

(McAnarney, 1985). However it is possible that maternal child rearing behaviours are implicated here, with these relationships. With higher expectations of her child a mother may be more proactive in the encouragement of autonomous behaviour and the development of self-help and adaptive skills. She may also be more knowledgeable about child development and more proactive in terms of child rearing, thus, not employing a learned helplessness attributional style. This would support previous research, which has linked such a parenting style to a decrease in child cognitive function over time (Thompson et al., 2002) and would support the relationship found in the present study between child cognitive function and the everyday level of assistance required by the child. Thus, despite organic causes of learning disability, other parenting, environmental, and family factors can contribute to a child's development. Notably the relationship between child learned helplessness and cognitive functional decline, parental factors such as low expectations of the child, their own learned helplessness attributional style, over-protection, and low child autonomy and self-reliance are implicated; and in terms of parent expectations of the child, supported by the present study.

The results produced from the correlational analyses illustrate further relationships between maternal appraisals and coping abilities and patient functional status. All full and subscale IQs positively related to maternal perceptions of child health and to perceived impact that the child's condition has on the family. Research has demonstrated the importance of stress processing factors like appraisal to parents' adjustment to having a chronically ill child. In particular, the degree to which mothers' perceive the child's illness to impact on the family, which has been highlighted as a mediating process through which illness or functional severity may lead to parental psychological problems (e.g. Wallander et al., 1989; Ireys and Silver, 1996; Lustig et al., 1996). Indeed, the correlational analyses also highlighted relationships between patient IQ and maternal SOC and parent distress, the latter variable showing negative relationships to Performance and Total IQ. This perhaps indicates visual impairment and a lack of manual dexterity which may have implications for functional care strain experienced by the mother.

In summary, although there are potential links between child cognitive function post-BMT and a number of medical and organic factors associated with MPS IH, the



findings of the present study also implicate aspects of the mother, particularly her expectations of the child and the seniority of her present or previous job in patient cognitive development. Since previous research has linked child learned helplessness to child cognitive development, it is possible that an attitude of resourcefulness and self-reliance, together with proactive health management strategies, are implicated in this relationship. This notion is worthy of consideration when providing support for families affected by this condition, particularly those with young children, as appropriate advice can be given regarding particular elements of parenting that may improve cognitive function to some degree. Clearly there is a link between the child's age at engrafted transplant and subsequent cognitive function. However, outcomes continue to be varied, and affected by other events or circumstances. There is also argument that other forms of transplant, such as cord blood, facilitate quicker treatment, lessen the chance of GvHD, and thus optimise cognitive functional outcome (Staba et al., 2004). Nevertheless, despite the type of treatment and other possible contributors to child cognitive function, early diagnosis and treatment of MPS IH continue to be of utmost importance, not only in optimising cognitive function, but also as an endeavour to improve other consequences of the condition post-BMT.

When considering the education of children affected by MPS IH post-BMT, it is first and foremost a matter of personal choice for a family, whether their child attends mainstream school or a school that is specifically tailored towards special educational needs. However, as the present study illustrates, most of the children aged under-12 years currently attend mainstream school with classroom support; and the majority of parents are satisfied with their child's educational progress, social integration, and emotional well-being within school. Sixty percent of those currently in secondary school attend mainstream school, and just under-40% of those no longer in full time education attended mainstream school. As the findings illustrate, cognitive function is higher for those under 12 years of age, which may explain the larger percentage of them attending mainstream school. With regard to the young adults in this sample however, factors including the little knowledge parents and medical professionals had about BMT outcomes when their children were young, and lower parental expectations of child future achievements (including longevity), were likely to have influenced parents' choices regarding their children's education. This is equally

relevant to the younger children in this sample, although in the reverse, as more is now known about treatment outcomes, and socio-culturally more emphasis is put on an inclusive education system.

In terms of the adolescents currently in secondary school, it is important to note however, that one individual was moved out of mainstream education into special needs education at the secondary school level, for social and emotional reasons as much as for educational reasons. This was also the case for an individual during secondary school, who is now no longer in full-time education. In both cases the individuals were female and both became particularly withdrawn in the mainstream secondary school setting. This highlights the importance of psychosocial support within the education system, particularly for children and young people with physical disability and mild to moderate learning disability. As previously discussed, children and adolescents with disabilities can experience social and interpersonal difficulties at school (Wray et al., 2001), and such difficulties can mask learning ability and render children being denied adequate education in the mainstream system (Whittington et al., 2004). This is particularly pertinent for this patient group, as they can be withdrawn and introverted individuals, particularly in adolescence, which makes them vulnerable to social difficulties. Learning difficulties and repeated academic failure are also pertinent to the psychosocial development of this patient group; and learning support with an emphasis on promoting feelings of achievement and adequacy is essential for this group of individuals. Appropriate and sustained psychosocial and educational support is therefore imperative for this patient group, so that they can continue to progress and learn along with their peers throughout the entire education system.

## 7.4 Patient Internalising and Externalising Behaviour

The Internalising Problems composite of the Behaviour Assessment System for Children (BASC) (Reynolds and Kamphaus, 1998) comprises the Anxiety, Depression, and Somatisation scales of the measure. This section describes maternal reports of patient internalising and externalising problems. Thirty-nine mothers were included in the analysis. Five with children aged under-2½ years were excluded due to the age parameters of the measure. The normative mean for each of the scales and composite scores on the BASC is 50 with a standard deviation of 10. In comparison to normative data as set out in the BASC manual, Table 7-14 below shows the mean scores of Internalising Behaviour for this patient group to be within the normal range, with 28% falling outside of this range (13% above a score of 60). Scores below 40 and above 60 on this composite measure indicate a cause for concern.

**Table 7-14 Descriptive Statistics for Patient Internalising Problems Across Age Group**

Age Group	N	Min	Max	Mean	Std. Dev	% outside of normative range
2½ – 5 years	12	39.00	67.00	51.83	8.58	30%
6 – 11 years	15	27.00	79.00	49.73	13.44	30%
12 years +	12	38.00	58.00	47.00	7.29	17%
All Ages	39	27.00	79.00	49.54	10.34	28%

A one-way analysis of variance was carried out on the data, and no significant differences were found between the mean scores of Internalising Problems for each age group.

With regard to Externalising Problems, which include hyperactivity, aggression, and conduct problems, again in comparison to normative data, the mean scores for each of the age groups of the present patient sample fell within the normal range. However,

46% of the sample was found to fall outside of that range (28% above a score of 60). A one-way analysis of variance revealed no significant differences between the mean scores of Externalising Problems for each of the age groups, as shown in Table 7-15 below.

**Table 7-15 Descriptive Statistics for Patient Externalising Problems Across Age Group**

	N	Min	Max	Mean	Std. Dev	% outside of normative range
2 <sup>1</sup> / <sub>2</sub> – 5 years	12	35.00	77.00	56.83	11.64	50%
6 – 11 years	15	31.00	78.00	50.20	12.50	30%
12 years +	12	33.00	67.00	49.33	12.81	58%
All Ages	39	31.00	78.00	51.97	12.45	46%

#### **7.4.1. Predictive Association between Risk and Resistance Factors and Patient Internalising and Externalising Problems**

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient internalising problems than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Risk factors that were added into the model were physical functioning and the level of assistance required carrying out activities of daily living as measured by the MPS I Health Assessment Questionnaire, and cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests. Intrapersonal factors added into the model were patient mental health and self-esteem as measured by the Child Health Questionnaire, and patient adaptive skills as measured by the BASC Parent Form. Stress processing factors

added into the model were two measures of mothers' perception of child health as measured by the Child Health Questionnaire, maternal parenting stress as measured by the Parenting Stress Index, maternal SOC as measured by the Sense of Coherence Scale, maternal mental health as measured by the GHQ, and maternal spiritual beliefs as measured by the Beliefs and Attitudes Scale developed for this study. Social-ecological factors that were added into the model were family cohesion as measured by the Child Health Questionnaire, active-recreational, organised, and controlled family environment as measured by the Family Environment Scale, maternal social support as measured by the Social Support Scale developed for this study, and family disagreements pertaining to support and understanding of the child as measured by the Family Discord measured developed for this study. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 7-16 below to show the relative contribution of each of the child and family dimensions to patient Internalising Problems.

**Table 7-16 Model for Patient Internalising Problems**

Variable	Standardised Beta	t-value	Sig
<b>F = 4.031<sub>2,12</sub>, p = .046 (95%)</b>			
Patient Physical Functioning	-.434	-2.828	.015*
Level of Assistance Required	.942	8.028	.000**
Mother's Perception of Child's Health	.456	4.309	.001**
Patient Cognitive Function	.428	7.382	.000**
Patient Mental Health	-.512	-6.516	.000**
Patient Self Esteem	-.258	-3.163	.008*
Patient Adaptive Skills	-.165	-1.531	.152
Maternal Parenting Stress	-.238	-2.666	.021*
Maternal Sense of Coherence	-.733	-8.650	.000**
Mother's Perceptions of Child Health 2	-.297	-3.398	.005*
Maternal Mental Health	.106	1.019	.328
Maternal Beliefs	-.309	-4.814	.000**
Family Cohesion	-.031	-.420	.682
Family Environment: Active			
Recreational	.160	2.412	.033*
Family Environment: Organisation	.157	2.985	.011*
Family Environment: Control	-.161	-1.975	.072
Support from Friends			
Family Disagreements: Immediate	.173	2.479	.029*
Family Support and Understanding of Child	-.177	-2.119	.056
* Significant at the 0.05 level			
** Significant at the 0.005 level			

The relationships found indicate that risk factors relating to functional status and level of assistance required relate significantly to patient internalising problems, with better cognitive and physical functioning and more assistance required relating to a higher incidence of internalising problems. Intrapersonal factors pertaining to patient mental health, self-esteem, and adaptive skills were also related to internalising problems, all showing negative relationships to this outcome variable. A number of stress processing factors also showed significant relationships to internalising problems, including mothers' perceptions of child health, maternal parenting stress and SOC, and maternal beliefs. The majority of these variables also showed a negative relationship to patient internalising problems. Social-ecological factors which were significantly related to patient internalising problems were an active-recreational and organised family environment, greater maternal social support, and fewer disagreements with family members regarding the support and understanding of the

child, although the relationship between this latter variable and that of patient internalising problems was of borderline significance. Ninety-five percent of the variance was explained by these factors, suggesting that a child's adjustment to living with this condition is influenced by illness-related factors and individual and family-related factors. These relationships will be discussed in terms of past and future research, and to practical applications.

Hierarchical multiple regression analyses were again employed to test the hypothesis that resistance factors will account for more of the variance in patient externalising problems than illness and disability related risk factors. The same risk and resistance factors were added into the model in the same order as above, with the exception of the social-ecological factors. For patient externalising problems an independence orientated family environment as measured by the Family Environment Scale, mothers and fathers' discrepant or shared coping abilities as measured by the Sense of Coherence scale, maternal social support as measured by the Social Support Scale developed for this study, and disappointment experienced by the mother regarding family lack of understanding regarding issues relating to the child's MPS condition were added into the model. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 7-17 below to show the relative contribution of each of the child and family dimensions to patient Externalising Problems.

**Table 7-17 Model for Patient Externalising Problems**

Variable	Stand. Beta	t-value	Sig
<b>F = 2.618<sub>2,9</sub>, p = .127 (91%)</b>			
Patient Physical Functioning	-.030	-.156	.880
Level of Assistance Required	.973	5.127	.001**
Mother's Perception of Child's Health	.047	.335	.746
Patient Cognitive Function	-.381	-3.664	.005*
Patient Mental Health	.044	.360	.727
Patient Self Esteem	-.187	-1.180	.268
Patient Adaptive Skills	.194	1.588	.147
Maternal Parenting Stress	-.460	-3.105	.013*
Maternal Sense of Coherence	.383	2.498	.034*
Maternal Mental Health	-.517	-3.375	.008*
Maternal Beliefs	-.549	-6.022	.000**
Family Environment: Independence	-.803	-7.007	.000**
Discrepant Reports of Parenting Stress	.530	2.875	.018*
Discrepant SOC scores between Parents	-.137	-.893	.395
Support from Friends	.086	1.056	.319
Disappointment: Issues Relating to Child's MPS Condition	.201	1.932	.085

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

The relationships found indicate that aspects of the patient's condition in terms of the level of assistance required carrying out activities of daily living, and other intrapersonal, stress processing, and social-ecological factors contribute to patient externalising problems, accounting for 91% of the variance. The intrapersonal factor of patient mental health showed a positive relationship to externalising problems, and two stress processing factors pertaining to maternal SOC, mental health, and spiritual beliefs showed both positive and negative relationships, indicating that externalising problems were greater when maternal SOC was higher, spiritual beliefs lower, and maternal mental health poorer. The social-ecological factors of family environment, particularly when it is independence orientated was negatively related to externalising problems, and discrepant coping abilities between mothers and fathers was positive related, indicating that externalising problems are higher when fathers' SOC was lower than mothers. These relationships will be discussed in terms of past and future research, and to practical applications.



#### **7.4.2. Discussion**

With regard to both internalising and externalising problems this patient group, overall do not appear to experience any greater or lesser difficulties than a normative population of children and young people. However, as with the general population, there is a range of scores for each of these composite measures, indicating that some children and young people affected by MPS IH do experience both internalising and externalising problems post-BMT. Indeed, the findings demonstrate that 13% of this patient population experience problems of an internalising nature and 28% of an externalising nature. With regard to internalising problems, over half of the variance of the multiple regression was explained by child intrapersonal characteristics, such as emotional well-being and self-esteem, and aspects of the MPS condition itself. More precisely, greater internalising problems were related to lower self-esteem, greater emotional difficulties, better health as perceived by the mother, better cognitive function, fewer physical disabilities, and a greater level of assistance required with activities of daily living. This indicates that internalising problems are experienced more by the patient participants who are perceived as being physically healthy and borderline physically and mentally able, but who also experience emotional difficulties and low self-esteem. Thus for individuals who have borderline functional competence both physically and cognitively, intrapersonal factors such as self-esteem can buffer the effects of illness related stressors.

The children and young people in this patient group are affected by learning disability to varying degrees, from profound to mild. In terms of physical disability, again, the degree to which a child is affected varies from individual to individual. However, by adolescence physical disability becomes more apparent, mobility significantly decreases, and more reliance is put on mobility aids to get around school and the neighbourhood. With mild to moderate learning difficulties then, it is likely that a child will be aware of differences between themselves and others in terms of physical and learning ability. Indeed, having milder learning difficulties is likely to make adjustment to living with MPS IH post-BMT more difficult for a child than being more severely affected. This supports previous research, which has found feelings of academic failure to affect children with learning difficulties socially and emotionally

by impacting upon their self-esteem and self-concept (Dean, 1985; Hersh et al., 1996; Pihl and McLarnon, 1984). Previous research has also found children with learning and communication impairments to experience poor psychosocial adjustment (Witt et al., 2003), and those with lower adaptive and developmental competencies to experience adjustment problems (Hurtig and Park, 1989; Casey et al., 2000). Additionally, with physical disability, if physical functioning is partly intact and the child is able to participate in physical activities, limitations due to the condition, fatigue, and pain, may be more upsetting for a child in this situation than when their disability is more marked. Thus they may wish to participate in activities with peers and be seen as no different to the rest of the peer group. Previous research is inconclusive regarding the relationship between physical functioning and psychosocial adjustment however (Heller et al., 1985; Lavigne and Faier-Routman, 1993; Padur et al., 1995). In terms of self-concept though, young people with progressive physical disability have been reported as becoming increasingly socially withdrawn as physical status declines (Harper, 1983) and social marginalisation can cause distress (e.g. Barlow et al., 1998; LaGreca et al., 1999).

Thus, with MPS IH, which is a visibly disabling condition, and affects an individual's growth and the formation of bones and joints, a young person is greatly challenged in life as he or she copes living with this condition. Moreover, as has been illustrated here, such coping is likely to be further challenged when a child has mild to moderate learning difficulties, and when physical functioning is limited, but not profoundly so. Both aspects of the MPS condition have implications for an individual's self-concept and concept of adequacy in comparison to peers, which may be exacerbated when the individuals' overall health is also perceived by the mother as good. Furthermore, as has been previously written, intrapersonal factors such as self-esteem and self-concept are key predictors of healthy adjustment in children and young people with disabilities and chronic illness (Anderson et al., 1982; Kapp-Simon, 1986); and as supported by the present study, with lower self-esteem the child may be more vulnerable to emotional problems of an internalising nature, which are linked to the overall coping experience of living with the MPS condition.

As the results of the present study illustrate, better physical and cognitive functioning are related to patient internalising problems. However, the more assistance the patient

requires carrying out activities of daily living is also associated with such problems. This measure was completed by the mother and therefore may be more indicative of mothers' perception of child need as opposed to an objective measure of child need. In this case it is possible that this result reflects a relationship between maternal over-involvement and perceptions of child vulnerability and child internalisation rather than between internalisation and child care needs directly. It may also reflect a learned helplessness response to the condition, where the individual over time withdraws from using self-help skills, or fails to develop them, in favour of reliance on others for assistance or withdrawal from the activity altogether.

Thus, both ideas would support previous research, which has linked parent cognitions like perceived child vulnerability to adjustment difficulties for the child including internalising problems (Estroff et al., 1994), increased social anxiety (Anthony et al., 2003) and an increase in child dependence, demandingness, and behavioural difficulties (Bendell et al., 1994). Higher perceived child vulnerability has also been related to more negative outcomes for the child in terms of adaptive functioning (Allen et al., 2004) and the interference of the development of autonomous behaviour in childhood (Anderson and Coyne, 1991; Holmbeck et al., 2002). It also supports previous research which has described learned helplessness in relation to psychosocial difficulties (Braden, 1990). This relationship therefore highlights the importance of autonomous behaviour and the development of resourceful and self-help skills to child psychosocial adjustment to living with disability and chronic illness (Braden, 1990); and implicates the role that parents can play in exacerbating learned helplessness behaviour.

Previous research has found mothers of children with disabilities to see their role as mother and carer as a priority, and their emotional well-being appears to be related to their satisfaction with their abilities within those roles (Pelchat et al., 2003). Maternal distress and adjustment have also been related to child psychosocial adjustment (Sawyer et al, 1998; Singer et al., 2003; Thompson et al., 2002). In support of this, the findings of the present study illustrate how stress processing factors relating to maternal coping in terms of Sense of Coherence is negatively related to child internalising problems. However, mothers' parenting stress also has a negative relationship with this outcome variable, which does not support these findings. In the

present study then, low maternal parenting stress is related to greater child internalisation. This suggests that the mother feels satisfied in her role as mother, although her coping resources are poor. In this instance the direction of the relationship between maternal parenting stress and child internalisation may indicate maternal over-involvement. However, if over-involvement in the care of the child is apparent it can interrupt the development of autonomy for the child as previously discussed. Externalising as well as internalising behaviours have been implicated in such parenting, as well as social withdrawal and lowered self-esteem (Anderson and Coyne, 1991; Holmbeck et al., 2002). Child Internalising Problems in the present study are also positively related to mothers' perception that the child is vulnerable in terms of their physical health, another stress processing factor. This relationship may, again, be indicative of maternal over-protection and perceptions of child vulnerability, and reinforces the relationships found between child internalisation and maternal parenting stress and mothers' rating of the level of assistance required by the child.

Finally, in terms of the stress processing factor of maternal spiritual beliefs, this variable and patient internalisation was also negatively related. Again, this is possibly indicative of poor maternal coping and an inability to ascribe positive meaning to the child's condition, which in turn is related to greater patient internalisation. Previous research has found parents who are able to give special and positive meaning to a child's illness to be better able to cope and adjust (McCubbin and McCubbin, 1993; Venters, 1981), and family stress processing and other resistance factors have been seen to function as stress processing factors relevant to children's adaptation (e.g. Burlew et al., 2000). Thus, in terms of maternal stress processing factors, poor coping, the perception that the child's health is poor, few beliefs in things spiritual, and low maternal stress are all related to greater child internalisation.

With regard to social-ecological factors relating to family environment, the results of the present study are difficult to interpret, as the direction of the relationships that have emerged are more often associated with positive child psychosocial outcomes. Here patient internalising problems are associated with a family environment that is active-recreational and organisation orientated, and low control orientated. Furthermore, patient internalising problems are related to greater support from friends for the mother and fewer disagreements with immediate family members about the

child and aspects of the child's MPS condition. One possible explanation for these relationships is that an increase in structure is often found in families of children with disabilities, which depicts organisation orientation, and is seen as an effective coping strategy (Moos and Moos, 2002). Families who exercise a stable routine are often high in organisation and control, low in conflict, and high in cohesion orientation (Fiese and Kline, 1993). However, when the emphasis on structure is high, it is possibly reflective of a pre-existing family characteristic that may contribute to a child's problems (Moos and Moos, 2002).

Another explanation is that, while family environment can have an impact on child psychosocial development, so child characteristics, disabilities, and dispositions can have an impact on, and alter, family environment (Moos and Moos, 2002). It is possible that, with the present patient population, when a child is more internalising the family accommodates it by adapting to it. They become more organised and structured, they partake in more family-orientated social activities, the child virtually becomes the centre of the family, and thus a cohesive family environment is formed around the child. Such a family environment could also be seen as protective when a child has the tendency to internalise. Organisation and the use of routines can be used to ensure that the child feels safe and secure, and recreational activities that revolve around the family can also be seen as a parentally controlled social environment, that may not in fact be appropriate for the child. In terms of social support, families who are more cohesive also see themselves as receiving more social support (Sarason et al., 1987). Furthermore, families orientated towards recreational pursuits are reported to have more friends and acquaintances (Moos and Moos, 2002), though again, such acquaintances may have been hand picked to fit in with the family environment that has been created around the child.

In summary then, patient internalising problems when the individual is affected by MPS IH post-BMT are related to aspects of the condition itself in terms of cognitive and physical functioning, particularly when functioning is moderate, and to intrapersonal characteristics of the child, particularly emotional well-being and self-esteem. However, such problems are also related to maternal stress processing factors, namely maternal parenting stress and coping, her spiritual beliefs, and perceptions of child vulnerability, and to social-ecological factors relating to family environment.

These findings have implications for the mothers' maternal role and possible over-involvement and perceptions of child vulnerability, and to aspects of the family. Since a large percentage of the variance was explained by aspects of the child and the MPS condition, it is clear that coping with living with MPS IH post-BMT is a challenge for children and young people, particularly when their learning difficulties are mild to moderate. Psychological support is therefore imperative for these children from a young age. Interventions, which concentrate on the development of autonomy, and subsequently self-esteem, are also important. However, such interventions are important within the home as well as within a learning environment such as school or nursery. It is important that children affected by MPS IH who have undergone bone marrow transplant, and who have mild to moderate learning difficulties are afforded autonomy, independence, and inclusion in education and social activities, inside and outside of school. Of course, all children affected by this disorder should be afforded such things post-BMT, however the findings of the present study illustrate the emotional impact the disorder can have on children when they have milder learning difficulties. The relevance of autonomy and self-esteem in relation to psychosocial adjustment post-BMT for these children needs to be disseminated to parents, educators, and health care providers alike, so that appropriate support can be given and outcomes improved.

In terms of patient externalising problems, the relationships that emerged from the present study appear to implicate more severe learning difficulties in this type of behaviour. Stress processing factors relating to mothers' parenting stress, coping, and beliefs and attitudes, indicate an acceptance of the child's condition and the behaviours that are demonstrated as a result. They imply the presence of a realistic view, that mothers do not suffer negative consequences as a result of the child's challenging behaviour, as though with such strong coping resources, mothers can tolerate and allow externalisation to continue. The relationship between a family environment that is independence orientated, also implies that the family accepts the child for who they are and gives the child the right to express themselves in their way. They accept the child and the child's condition and continue to fulfil their individual needs in life. Such externalising behaviour is possibly most apparent however, around other people, in particular members of the extended family. The results illustrate how mothers' perception that members of the extended family do not truly understand the

child's condition and the BMT treatment, albeit unexpressed feelings, possibly highlight externalising behavioural problems for the mother. Parents of children affected by MPS IH post-BMT who have more severe learning disabilities would benefit from interventions which enable the effective management of externalising behaviours, together with appropriate education programmes, and respite care if necessary. Research has found behaviour management programmes and psycho-education interventions for mothers of children with autism to greatly benefit them by essentially raising their Sense of Coherence *comprehensibility* and *manageability* (Bristol et al., 1993; Olsson and Hwang, 2002). Time and space for mothers to explore their own goals would also be beneficial to their well-being and in turn to the efficacy of their parenting (Breslau et al., 1982). Being able to redefine a child's disability as meaningful and positive can enhance well-being in mothers, which in turn increases acceptability of the child and the child's condition, and in turn again improves parenting (Soloman et al, 2001).

## 7.5 Patient Self-Image

Twenty-two patient participants were included in this analysis, which describes patient scores on the Butler Self-Image Profiles (2002). Independent samples t-tests were employed to discover whether patient participants scored differently according to their age. No significant differences were found between patient participants aged 7-11 years and those aged 12 years and over in terms of Positive Self-Image, Negative Self-Image, and Sense of Difference. Descriptive statistics are shown in Table 7-18 below.

**Table 7-18 Descriptive Statistics for Patient Positive Self Image, Negative Self Image, and Sense of Difference**

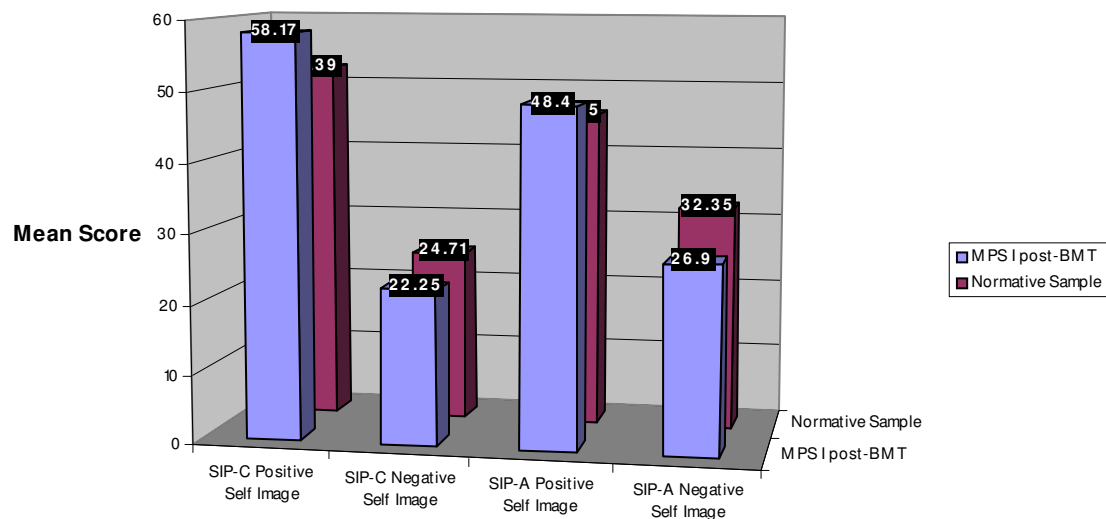
	N	Min	Max	Mean	Std. Dev
<b>Positive Self Image</b>					
7-11 years	12	33.00	72.00	58.17	12.71
12 years +	10	25.00	67.00	48.40	14.09
<b>Negative Self Image</b>					
7-11 years	12	6.00	48.00	22.25	13.88
12 years +	10	6.00	51.00	26.90	16.57
<b>Sense of Difference</b>					
7-11 years	12	1.00	6.00	4.00	2.00
12 years +	10	.00	6.00	2.50	2.32

In terms of normative data, mean scores and standard deviations are given for each year group by the author of the measure (Butler, 2001). However, since the sample size of the present study is so small, averages of the mean scores given for the 7-11 year age group and the 12 years and over age group were calculated and used. In terms of Positive and Negative Self-Image then, in comparison to normative means, the mean scores of the individuals in both age groups did not differ significantly from those of the normative sample, as shown in Figure 7-8 below. Interestingly, the mean



scores of the MPS IH patients followed the same pattern as those of the individuals in the normative sample. The older group generally saw themselves as less positive and more negative than the younger group.

**Figure 7-8 Self Image Profiles for Children (7-11 years) and Adolescents/Young Adults (12 + years) Positive and Negative Self Image in Comparison to Normative Data**



### 7.5.1. Predictive Association between Risk and Resistance Factors and Patient Positive and Negative Self-Image

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient positive self image than illness and disability related risk factors. Variables were entered into the hierarchy in theoretical groups as follows: 1) illness-related risk factors, 2) intrapersonal resistance factors, 3) stress processing resistance factors, and 4) social ecological resistance factors. Risk factors that were added into the model were physical functioning and the level of assistance required carrying out activities of daily living as measured by the MPS I Health Assessment Questionnaire, and

cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests. Intrapersonal factors added into the model were patient mental health and self-esteem as measured by the Child Health Questionnaire, and patient adaptive skills as measured by the BASC Parent Form. Stress processing factors added into the model were two measures of mothers' perception of child health as measured by the Child Health Questionnaire, maternal parent distress as measured by the Parenting Stress Index, and maternal SOC *comprehensibility* as measured by the Sense of Coherence Scale. Social-ecological factors that were added into the model were intellectual-cultural family environment, independence, and family cohesion as measured by the Family Environment Scale. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 7-19 below to show the relative contribution of each of the child and family dimensions to child and adolescent Positive Self-Image.

**Table 7-19 Model for Patient Positive Self-Image**

Variable	Stand. Beta	t-value	Sig
<b>F = 2.951<sub>3,6</sub>, p = .120 (76%)</b>			
Patient Physical Functioning	-.270	-1.089	.318
Level of Assistance Required	.786	3.232	.018*
Mother's Perception of Child's Health	-.516	-2.724	.034*
Patient Cognitive Function	.097	.477	.650
Patient Self Esteem	.382	1.081	.321
Patient Adaptive Skills	.322	1.253	.257
Patient Mental Health	.192	.664	.531
Maternal Beliefs	-.154	-.730	.493
Maternal Parent Distress	-.921	-4.003	.007*
Maternal SOC Comprehensibility	-.940	-4.020	.007*
Maternal Perceptions of Child Health 2	.022	.129	.901
Family Environment: Intellectual-Cultural	.451	2.640	.039*
Family Environment: Independence	.107	.545	.606
Family Environment: Cohesion	-.256	-1.335	.230

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

With all the variables entered into the analysis, risk factors related to the assistance the patient requires carrying out activities of daily living, stress processing factors relating to mothers' perception of child health, parenting stress and coping, and

social-ecological factors relating to family environment contributed to patient positive self-image, accounting for 76% of the variance. Positive self-image appears to be positively related to the level of assistance required by the individual carrying out activities of daily living and negatively related to mothers' perception of overall patient health. It is also negatively related to maternal parent distress and SOC *comprehensibility*, and positively related to a family environment that is intellectual cultural orientated. These relationships will be discussed in relation to previous and future research, and to practical applications.

Hierarchical multiple regression analyses were employed to test the hypothesis that resistance factors will account for more of the variance in patient negative self-image than illness and disability related risk factors. Risk factors that were added into the model were physical functioning and the level of assistance required carrying out activities of daily living as measured by the MPS I Health Assessment Questionnaire, and cognitive function as measured by the Griffiths, WISC-III, and WAIS-III developmental tests. Intrapersonal factors added into the model were patient mental health and self-esteem as measured by the Child Health Questionnaire, and patient adaptive skills as measured by the BASC Parent Form. Stress processing factors added into the model were two measures of mothers' perception of child health as measured by the Child Health Questionnaire, maternal spiritual beliefs as measured by the Beliefs and Attitudes scale developed for this study, maternal parent distress as measured by the Parenting Stress Index, and maternal SOC *comprehensibility* as measured by the Sense of Coherence Scale. Social-ecological factors that were added into the model were intellectual-cultural family environment, independence and cohesion as measured by the Family Environment Scale. The standardised regression coefficients, t-values, and probabilities for the final model are reported in Table 7-19 below to show the relative contribution of each of the child and family dimensions to patient Negative Self-Image.

**Table 7-20 Model for Patient Negative Self Image**

Variable	Stand. Beta	t-value	Sig
<b>F = 2.973<sub>3,6</sub>, p = .119 (79%)</b>			
Patient Physical Functioning	-.590	-2.497	.047*
Level of Assistance Required	-.014	-.060	.954
Mother's Perception of Child's Health	-.051	-.285	.785
Patient Cognitive Function	.334	1.730	.134
Patient Self Esteem	.945	2.813	.031*
Patient Adaptive Skills	-1.082	-4.429	.004**
Patient Mental Health	-.357	-1.299	.242
Maternal Beliefs	-.023	-.113	.914
Maternal Parent Distress	-.412	-1.882	.109
Maternal SOC Comprehensibility	-.178	-.799	.455
Maternal Perceptions of Child Health	-.171	-1.073	.324
Family Environment: Intellectual-Cultural	.394	2.426	.051
Family Environment: Independence	-.261	-1.393	.213
Family Environment: Cohesion	.140	.771	.470

\* Significant at the 0.05 level

\*\* Significant at the 0.005 level

With all the variables entered into the analysis, a disability related risk factor and intrapersonal and social-ecological resistance factors contributed to patient negative self-image, explaining 79% of the variance. Negative self-image appears to be greater when physical functioning is less impaired, adaptive skills less well developed, and self-esteem higher. It is also positively related to a family environment that is intellectual-cultural orientated, although this relationship is of borderline significance. These relationships will be discussed in relation to previous and future research, and to practical applications.

### 7.5.2. Discussion

Overall, the children and young people in this sample did not see themselves any more negatively or any less positively than the individuals in the normative sample. This is supported by previous research of adolescents with physical disability who do not see themselves as different from their healthy peers and do not see themselves in a less positive light than their non-disabled peers (Skar, 2003). With regard to positive self-image then, the results of the present study illustrate how that of the children and

young people affected by MPS IH post-BMT is related to aspects of the disease in terms of the level of assistance required with activities of daily living, to maternal stress processing factors relating to maternal stress and coping, and perceptions of child health, and to social-ecological factors relating to family environment.

In terms of the health and disability factors that contributed to patient positive self-image both measures were completed by the mother. One measured mother's perception of child health and therefore constitutes a stress processing factor. The other measured the level of assistance the child required carrying out activities of daily living. Thus it is the mothers' perception of the child's health and assistance requirements that is highlighted here; and positive self-image may therefore be related to the amount of attention the child receives from the mother. Although previous research has found adolescents with disabilities to feel their lives to be over-controlled by adults (Skar, 2003) and maternal perceptions of child vulnerability to impact on child psychosocial adjustment (e.g. Anthony et al., 2003), the very role of being 'sick' or 'disabled' is purported as being a social role that can have advantages (Parsons, 1951), and to form a crucial part of one's identity when a condition is chronic and life-long (Royer, 1998). It is also possible that the relationships between these two variables and positive self-image are indicative of family support, which has been associated with the child psychosocial adjustment (Nihira et al., 1980; Wallander and Varni, 1989; Varni et al., 1991), and predictive of feelings of self-worth in young people with physical disabilities (Antle, 2004). Furthermore, although all children and young people affected by MPS IH will receive a lot of attention from medical professionals in their lives post-BMT, perhaps with poorer health and more pronounced orthopaedic difficulties such attention would be greater. Such attention would become a normal part of the individual's life and could indeed reinforce the child's perception that he or she is special. Such bolstering of a positive self within the sick role would continue to take place within the part of the child's world that is family/illness-bound however.

The idea that patient positive self-image stems from the family and from the child's identity with their sick role is possibly supported by the maternal stress processing factors that are implicated here, as low maternal parent distress and poor coping abilities (low SOC) are related to child positive self-image. With regard to maternal

parent distress, this factor could be viewed as mothers' adjustment to having a child with MPS IH post-BMT, which in particular has been associated in previous research of maternal adjustment to child disability with child psychosocial adjustment (Lavigne & Faier-Routman, 1993). Moreover, in the present study mothers' satisfaction in her maternal role is related to a more positive child self-image. As previously discussed, research has found mothers' well-being to be related to her satisfaction with her abilities as a mother (Pelchat et al., 2003). This suggests then, that greater maternal parenting satisfaction, and thus improved maternal well-being, is perhaps related to more attention, care, and nurturing behaviours being directed towards the child. With regard to maternal SOC *comprehensibility* however, this relationship is difficult to interpret, as one would expect child positive self-image to be positively related to maternal SOC. However, in the present study, a negative relationship has emerged between the two variables, suggesting that the less structured, predictable, and explicable the mother finds the world, the more positively the child sees him or herself. It is possible that this result indicates a maternal attitude that prioritises the child's quality of life and healthy adjustment, despite living in an unpredictable and incomprehensible situation. Thus, the emphasis for the mother is perhaps on the child's enjoyment of life however long or short it is perceived to be. Although unable to have a clear picture of the child's future, it is possible that a child's positive self-image is the result of a mother's focus on the child's adaptation with a positive 'lust for life' attitude. Indeed, families of children with disabilities have difficulty conceiving of the future, preferring to live 'one day at a time' (Ray, 2002), which this theory would reflect. This approach has however been conceptualised as an intrapersonal coping strategy, which allows a parent to gain mastery and control over their situation (e.g. Atkin and Ahmad, 2000) and which protects against engulfment (Twigg and Atkin, 1994). If the child's positive self-image stems from the family environment, and that is dominated by maternal nurturance, the more emphasis the mother puts on the child's adaptation and welfare the more positive the child would see him or herself.

In terms of the social-ecological factors relating to family environment, the findings of the present study show a positive relationship between an intellectual-cultural orientated family environment and child positive self-image. Previous research has found a family environment that is intellectual-cultural orientated to be related to

higher parental Sense of Coherence and more satisfaction with their families (Margalit et al., 1988), and to better school adjustment among children with learning disability (Nihira et al., 1981). It may be possible that an intellectual-cultural orientated family enables a child to intellectualise aspects of themselves and their lives more readily, and to be able to formulate a view of him or herself. Interestingly however, in terms of positive self-image, this seems to be more the case for the children and young people over the age of 12, as the two variables were positively correlated for this age group, but not for the younger age group. This suggests that the ability to objectify oneself in a positive light when living with MPS IH post-BMT perhaps comes as the child grows into adolescence, a time when a child has a clearer idea of body function and illness causality (Lewis & Vitulano, 2003).

Interestingly, a family environment that is intellectual-cultural orientated is also positively related to negative self-image in the present study. Thus, it is possible that for the present sample that completed this measure an intellectual-cultural orientated family environment enables a child or young person to view him or herself in both a positive and negative light, but that intellectual-cultural orientation facilitates a more positive self-image when the individual is in adolescence. Continuing with negative self-image then, the other variables to contribute include patient physical functioning, self-esteem and adaptive skills. More precisely, negative self-image was related to better physical functioning, better self-esteem, and less well-developed adaptive skills. If one views the positive self-image of this patient group as being related to the safe and enclosed family environment that is also ensconced in the sick role, it is possible to view the negative self-image in terms of the child or young person's social world outside of the family. Better physical functioning may indicate a borderline state of physical ability in terms of participation in activities with peers. A greater negative self-image may relate to the child's limitations in terms of physical ability, which is exacerbated when the child is partly able to participate but cannot fully keep up with peers. This supports previous research, which has reported physical functioning and the ability to carry out age appropriate tasks to have an impact on child psychosocial adjustment when they are affected by disability (Heller et al., 1985), and perhaps indicates a lowered perceived athletic competence (King et al., 1993). It may also support research, which has found adolescents with physical disabilities to feel their peers to view them as different (Skar, 2003).

Similarly the relationship between adaptive skills and negative self-image may highlight limitations in the child's social abilities and their exclusion from the peer group and peer group activities. Adaptive skills measure social and communication skills, which if lacking can have an impact on children's social competence. It is possible that the lack of inclusion is extended to social activities that are not necessarily physically active in nature. Previous research has found young people with physical disabilities to have lower perceived social acceptance (King et al., 1993), and for those with learning difficulties to have low self-concept and social difficulties (Pihl & McLarnon, 1984). It has also found a significant association between adaptive behaviour and maladjustment (e.g. Casey et al., 2000). Finally, child self-esteem was positively related to child negative self-image, which is an interesting finding. A possible explanation for this is general rambunctiousness during interview, with children being quite cheeky and full of themselves, feeling able to say negative things about themselves as a way of showing off. Moreover, a child with a higher self-esteem may be more able to say negative things about themselves than a child with a lower self-esteem as a certain amount of confidence and courage is required. It is also worth noting that all child interviews were conducted in the family home, the place where the children were more likely to feel most confident. Interviews were carried out in private in a one-to-one researcher-participant environment within the home. This, in itself, may have instilled more confidence in the individuals.

It is possible then that the findings of the present study highlight the importance of the family environment to the development of positive self-image, but the importance of the social environment to the development of negative self-image. Thus, illustrating the importance of adaptive skills to an individual's self-esteem and social competence, and raising the appropriateness of certain parenting strategies to the development of such skills. Although aspects of caring for and nurturing a child with MPS IH post-BMT may develop positive self-image within the confines of the home, the hospital and the family, whether that translates to the child's social world that is outside of the family is called into question here. The findings of the present study highlight the importance of the development of autonomy, independence, and a sense of self from within a loving and supportive family environment, which equips a child



with adequate adaptive skills so that he or she can develop the internal coping resources to be socially competent in the outside world. Organisations, which provide support to families affected by MPS IH post-BMT, must therefore consider parenting and family factors more directly when considering the quality of life and psychosocial adjustment of the children. The spectre of death, which looms over these families is an issue that requires immediate attention. If families are to parent their children with a future in mind, it is important that doubts about longevity post-BMT are allayed as much as possible by the medical profession. Issues of parental over-protectiveness are valid for these parents, particularly mothers, which are understandable in the light of the devastating diagnosis and uncertainties following treatment. However, if child psychosocial outcomes are to be improved, parenting issues such as these require attention.

## **7.6. Summary of Findings: Patient Participant Adjustment Outcomes**

The aims and objectives of the study relevant to this section of the analysis were to examine the moderating effects of psychosocial resistance factors on the relationship between illness-related stressors and patient adjustment. It was hypothesised that patient participants would have significantly less well developed adaptive and social skills than a normative sample and that they would exhibit behavioural problems of an internalising nature. It was also hypothesised that psychosocial resistance factors, including intrapersonal, stress processing, and social-ecological factors, would contribute to the variance in patient adjustment and that they would moderate the negative effects of illness-related stressors on adjustment outcomes.

The children and young people affected by MPS IH post-BMT that participated in this study were found to have adaptive and social skills that were not well developed in comparison to a normative sample. This was particularly the case for the younger ( $2\frac{1}{2}$  – 5 years) and older (12 years plus) individuals. These findings therefore partially support the hypothesis that individuals affected by this condition would have less well developed social and adaptive skills than a normative sample. The factors that were found to make a significant contribution to social competence outcomes included intrapersonal (self-esteem, adaptive skills), stress processing (maternal parenting stress and coping (SOC), maternal perceptions of child health) and social-ecological factors (family environment). Maternal perceptions of child health and anxious cognitions about child welfare were highlighted as stress processing factors that contributed to patient adaptive skills in particular. Cognitive function was an illness-related risk factor that was significantly positively related to patient social skills. Thus, while intrapersonal, stress processing, and social-ecological factors contributed to the variance in patient social competency (78% Adaptive Skills, 77% Withdrawal, 86% Social Skills), functional independence played an important role in one aspect of this, social skills. Illness-related and psychosocial risk factors pertaining to cognitive function and the level of assistance the individual required carrying out activities of daily living were also found to be significantly related to patient personal adjustment. Other intrapersonal (adaptive skills), stress processing (maternal SOC), and family

environment factors were also found to significantly predict this outcome. These variables accounted for 95% of the overall variance in patient personal adjustment.

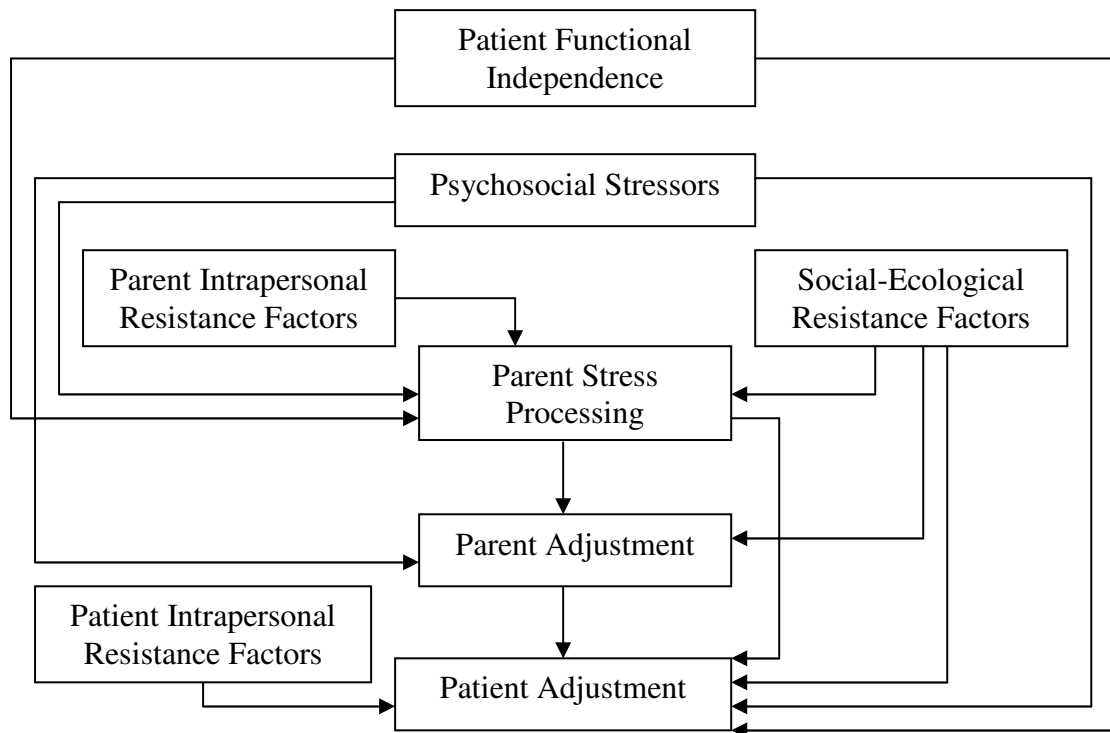
Similarly with patient internalising problems, a number of illness-related and psychosocial risk factors were found to significantly predict this outcome. These related to physical and cognitive functioning, and the level of assistance the individual required carrying out activities of daily living. However, a number of intrapersonal (patient mental health, self-esteem), stress processing (mothers' perception of child health, mothers' stress and coping, maternal spiritual and fatalistic beliefs), and social-ecological (family environment, mothers' social support) factors were also found to significantly contribute to patient internalising problems, accounting for 95% of the overall variance. Patient externalising problems were also found to be significantly predicted by a combination of illness-related and psychosocial risk factors (cognitive function, level of assistance required), stress processing (maternal stress and coping, mothers' mental health, mothers' spiritual and fatalistic beliefs), and social-ecological (family environment) factors, accounting for 91% of the overall variance. Overall, this patient population was not found to exhibit behavioural problems of an internalising, thus the hypothesis in relation to this was not supported. However, 13% of this patient population were found to experience internalising problems and 28% externalising problems, which should be noted for future research and patient support.

Patient positive self-image was significantly predicted by a psychosocial risk factor which referred to the level of assistance the individual required carrying out activities of daily living. However, it was also significantly predicted by stress processing (maternal perceptions of child health, maternal parent distress and SOC comprehensibility), and social-ecological (family environment) factors. These variables accounted for 76% of the overall variance. Negative self-image on the other hand was found to be significantly predicted by patient intrapersonal factors pertaining to self-esteem and adaptive skills, and to an illness-related risk factor relating to patient physical functioning. These variables accounted for 79% of the overall variance in patient negative self-image.

In terms of cognitive and adaptive function, patient participants showed significant developmental delay and cognitive function impairment. The range of ability was wide however, and the subscales on all of the developmental and cognitive function tests utilised demonstrated how patient participants were particularly hindered in their completion of performance items due to orthopaedic disability and visual impairment. In terms of the predictors of patient cognitive functioning two psychosocial risk factors showed significant relationships. These were the number of visits the individual had made to their specialist MPS hospital in the previous 12 months and the level of assistance the individual required carrying out activities of daily living. In addition to this however, a stress processing factor related to mothers' expectations of the child, and a social-ecological factor relating to family utilitarian resources (mother's job classification) also significantly predicted this outcome.

These findings indicate how illness-related and psychosocial risk factors can impact directly on patient psychosocial adjustment. These factors particularly pertained to functional independence in terms of cognitive and adaptive functioning, and to the psychosocial risk factor relating to the level of assistance the individual required from parents carrying out activities of daily living. For the most part however, these findings demonstrate the importance of intrapersonal resistance factors, namely self-esteem and adaptive skills, in moderating the relationship between illness-related stressors and adaptation. They also highlight the role that parent stress processing factors, like parent appraisals, perceptions of child vulnerability, and parenting stress and coping, play in patient adjustment, and the moderating role that social-ecological factors relating to the family environment, a shared view of parenting between mothers and fathers, and maternal social support play in buffering the negative effects of illness-related stress on patient adjustment. Overall then, these findings support the hypothesis that psychosocial resistance factors will significantly contribute to patient adjustment outcomes and that they will moderate the negative effects of illness-related stressors on patient adjustment. These relationships are illustrated in Figure 7-9 below. The model supports the risk-resistance framework presented by Wallander and colleagues (e.g. 1989; 1992) as it demonstrates direct relationships between illness-related and psychosocial risk factors and patient adjustment. It also demonstrates the moderating effect of psychosocial resistance factors on the relationship between these risk factors and patient psychosocial adjustment.

**Figure 7-9. Model for Patient Adjustment**



## **CHAPTER EIGHT**

### **CONCLUSIONS**

The study had two main aims. The first was to explore the experiences of families affected by MPS IH post-BMT in an initial exploratory phase using qualitative analysis. The second was to examine the moderating effects of resistance factors on the effects of psychosocial and illness-related risk factors on parent and child adjustment in a second (quantitative) phase. The objectives of the project as a whole were four-fold: Firstly, to explore the experiences of parents of children and young people affected by MPS IH post-BMT, identifying the rewards and stresses associated with the experience and the factors that moderate the effects of stress on parent adjustment. Second and thirdly, to examine the degree to which intrapersonal, social-ecological, and stress processing resistance factors moderate the effects of psychosocial and illness-related risk factors on parent and patient adjustment. Finally, to assess whether spouses differentially experience parenting stress, coping, and perceptions of child care needs, and whether potential discrepancies in scores or a shared view has implications for parent well-being, and in turn, child psychosocial development by serving as a moderating factor.

The first qualitative phase highlighted a number of issues pertinent to parents of children affected by this condition, including the devastating experience of receiving the diagnosis of MPS I for one's child and feelings of uncertainty regarding the BMT, further corrective treatments, and the child's future physical, adaptive, and psychosocial functioning, among others issues. The importance of parent appraisals was also highlighted in relation to coping. This was evident in parents' accounts of their perceptions of sources of stress and support, the way in which they perceived their child's condition and abilities, the meaning they ascribed to the illness and their child's presence in the world, and their perceptions of child vulnerability. The potential impact these appraisals can have on parenting and on child adjustment was also highlighted. The findings from this qualitative phase supported and informed the second phase of the study by illuminating risk (psychosocial and illness-related) and resistance (intrapersonal, stress processing, and social-ecological) factors, which could be examined in relation adjustment outcomes. The second phase therefore

investigated the moderating effects of resistance factors on parent and patient psychosocial adjustment. Parent outcomes examined in the second phase included mothers' and fathers' parenting stress and coping, mothers' mental health, her expectations of the child and anxious perceptions about child welfare and risk. Patient participant outcomes examined included social competency, personal adjustment, cognitive and adaptive functioning, internalising and externalising behaviour, and self-image.

The moderating effect of psychosocial resistance factors on the relationship between illness-related stressors and both parent and patient adjustment was highlighted by this research. While functional independence was found to contribute to parent stress processing and patient adjustment, and psychosocial stressors to parent stress processing and parent and patient adjustment, social-ecological factors pertaining to the family environment and support, and stress processing factors associated with appraisals and coping strategies were also found to significantly contribute to parent adjustment outcomes. Moreover, this research demonstrated how intrapersonal factors such as self-esteem and adaptive skills, social-ecological family factors, and parent stress processing contribute to patient adjustment. Furthermore, it demonstrated how discrepant reports of parenting stress and of the child's care needs between mothers and fathers served as a social-ecological resistance factor and contributed to parent and patient adjustment. The aims and objectives of this study were therefore fully met, and the hypotheses set addressed and discussed in the last chapter. Since psychosocial resistance factors are amenable to change, it is important that the factors that contribute to adjustment are considered when developing intervention strategies designed to improve patient outcomes. In future research and in providing support to individuals and their families, it is therefore important to consider the whole family.

## **8.0. Recommendations for Future Research and Practical Applications**

In terms of parent stress and coping, previous research and the findings of the present study have highlighted the importance of family factors and spousal support for both mothers and fathers. They also highlight how mothers and fathers can have different concerns regarding the care and welfare of their child. For mothers parenting stress may be related to the commitment to care given from the child's infancy into adulthood. Indeed, this issue was reflected in the findings of the qualitative phase of this study by mothers of older children as a source of stress and in the analysis of sense of coherence in the second phase of the study. While the family environment and issues of family support were demonstrated as being important for both mothers' and fathers' coping, the level of assistance the child required carrying out activities of daily living was a psychosocial risk factor that further contributed to mothers' SOC. Stress processing factors relating to perceptions of child health and the impact the child's illness had on the family also contributed to mothers' SOC. Stress and coping, social and spousal support, perceptions of child health, and psychosocial stressors were also found to be related to mothers' mental health. These factors therefore require consideration by health and support professionals so that appropriate support can be given to families. By encouraging both parents to be involved in the medical and psychosocial support of the child, maternal engulfment in the caring role could be reduced. Play and parenting workshops when the child is in infancy could assist fathers in bonding with their child. Opportunities to communicate and participate in activities with other families who have children affected by the same condition, and which involve extended family members could also help mothers and fathers to feel more understood and less isolated by their situation. The experiences of fathers also require more in-depth study in future research. Exploration of the relationship between mothers' and fathers' involvement with the child in early infancy and their attitudes towards the parenting role, and their involvement and levels of parenting stress at later stages in the child's life would be useful.

The investigation of mothers' appraisals regarding their expectations of their child's future achievements highlighted child adaptive behaviour as a contributing factor. It



also highlighted that maternal stress and coping, family cohesion, family environment, and spousal support also contribute to this outcome. Similarly, mothers' anxiety about child welfare highlighted the child's adaptive and cognitive functioning as contributing factors plus a number of intrapersonal, stress processing, and social-ecological factors, including mothers' perceptions of child health. Mothers' perceptions of offspring risk were however solely predicted by social-ecological factors pertaining to the family environment. Since these outcomes relate essentially to stress processing and mothers' perceptions of child vulnerability, which in turn were demonstrated to be important to patient psychosocial adjustment, the factors that contribute to them require attention. Further research of a longitudinal nature would be useful so that relationships between parental concern for child welfare and perceptions of risk, parenting style, and child psychosocial and lifestyle outcomes can be explored. Future research of this nature would also benefit from the inclusion of all family members including fathers and siblings, so that family functioning, support, parental attitudes, and the role that siblings play in the reduction of maternal anxiety can be explored.

With regard to the provision of services to families affected by this disorder, it is important that families are given as clear an idea as possible about life span and physical and adaptive functioning following BMT. This would help to reduce feelings of uncertainty regarding the child's future functioning and indeed longevity, reducing in turn parent perceptions of child vulnerability. Further research regarding orthopaedic difficulties and the outcomes of corrective surgery would also be beneficial in reducing parental anxiety about making such decisions and about the child's susceptibility to injury. As previously mentioned, parenting workshops that involve both parents and other family members would have the advantage of promoting a greater understanding about the condition and the caring role, and facilitate a joint approach to parenting and the development of the father-child bond. The dissemination of parenting guidelines and the implications of parenting on child psychosocial outcomes might also assist families in looking to the child's future as well as living day by day.

The patient participants of this study in young childhood and adolescence/early adulthood were found to have under-developed social and adaptive skills and to be

prone to withdrawal. The older patient participants were also found to be socially inhibited, particularly in the school environment. In terms of internalising behaviour, 13% of the patient population were found to experience difficulties of this nature. These individuals were predominantly aged under-11 years. Since this study was cross-sectional no links could be made between social and adaptive functioning in early childhood and later adolescence and adulthood. Examination of patient social competency and internalising problems using a longitudinal design would therefore be useful in identifying children that may be at risk of adjustment disorders who are affected by this condition. This would aid the development of intervention, and family and child support programmes. Intrapersonal resistance factors related to self-esteem and adaptive skills were found to contribute to a number of these patient outcomes. In particular, self-esteem was found to contribute to patient adaptive and social skills, personal adjustment, and self-image, and to counter internalisation. Adaptive skills were found to contribute to social skills, personal adjustment, and self image. Further to this, a number of family environment and parent stress processing factors were found to contribute to patient adjustment outcomes.

As demonstrated by this and previous research, family functioning and support factors are essential to the development of a child's internal coping resources, and parent stress processing factors such as parent appraisals, perceptions of child vulnerability, and coping strategies serve as stress processing factors relevant to child adaptation on a number of levels. Therefore any support initiatives aimed at optimising child quality of life and improving child psychosocial outcomes should employ a family inclusive approach, and should include parent coping and adjustment in their considerations. For example, family support initiatives that promote child participation in group activities might enable young children to develop a greater sense of self-esteem and self-reliance and to foster adaptive skills, particularly after their time spent in semi-isolation following the BMT. Initiatives that employ a family-inclusive approach and encourage recreational activities and independence might also permit parents to feel more confident about their child's abilities and to be less over-protective, whilst simultaneously enabling children to develop social and adaptive skills. Thus, the role that parents play in facilitating the MPS child's social life would be a worthy topic for future research.

Support in the school environment is also essential for young people affected by this condition, so that a sense of adequacy, achievement, and self-esteem can be achieved from a young age throughout the individual's academic career. Functional independence and psychosocial stressors related to the child's condition were illustrated as factors that contributed to patient adjustment outcomes, particularly in terms of social skills and personal adjustment; and physical functioning was related to patient negative self-image. In these cases however, more negative adjustment outcomes were related to functional impairment that was mild to moderate. Social inhibition in relation to the school environment was also apparent for older individuals, suggesting a learned helplessness response to academic failure. Special attention therefore needs to be given to young people affected by MPS IH post-BMT as they enter secondary school, particularly in relation to appropriate classroom support and inclusion in classroom and social activities. Specific psychosocial support within the school environment should also be considered. Research of a longitudinal nature that examines learning disability and psychosocial development within the school environment would aid the development of appropriate support initiatives.

As well as academic support within the school, the promotion of autonomy and development of self-help skills can be facilitated in the home, which would optimise the psychosocial functioning of the child. The findings of the present study illustrated how maternal stress processing factors relating to her expectations of the child and the seniority of her present or previous job classification contributed to the child's cognitive functioning. Factors other than organic causes are therefore implicated in this relationship and are worthy of consideration when providing support to families, as appropriate advice can be given regarding elements of parenting that may improve cognitive functioning. Further research of this nature would also be useful in identifying the parenting factors that relate to functional outcomes for children affected by this condition and the role that they play in their development. In terms of the relationship between the child's age at transplant and subsequent cognitive function, a clear link was demonstrated. However, outcomes continue to be varied, and affected by other events or circumstances, including it is suggested the type of transplant undergone. Further research is needed regarding the effectiveness of different transplant types (cord blood, stem cell, and bone marrow) and regarding pre-

symptomatic transplant. This type of research should continue to be of the utmost importance, not only in optimising cognitive function, but also as an endeavour to improve other consequences of MPS IH post-BMT.

In conclusion then, since this is the first study to examine the experiences of this patient group and their families, it is essential that the findings are disseminated to service providers at the local level and to parents directly. This would include health care workers and counselling services that operate within the specialist hospitals that deal with MPS and related diseases, family support organisations, and child educators. Workshops and audiovisual and written materials aimed at parents would also be very useful in supporting families, particularly in the development of appropriate parenting strategies, as they adjust to having a child affected by MPS IH post-BMT. Such materials might also highlight to parents potential difficulties the child might face and possible ways of coping with them and improving psychosocial outcomes. Opportunities to meet other families of children affected by the same disorder might also assist families in adjusting to the diagnosis, having realistic expectations about the child's future, coping with potential problems, and optimising outcomes. Psychological support is also imperative for children and young people affected by this condition from a young age. Interventions aimed at developing autonomy, self-esteem, and social and adaptive skills would greatly benefit them. Such interventions are important as independent endeavours, as family-inclusive strategies, and within a learning environment such as school or nursery. This is particularly the case for those individuals affected by MPS IH post-BMT who have mild to moderate learning difficulties, so that their potential to experience adult social maturity and independence is maximised.

### **8.1. Strengths and Limitations of the Study**

The main limitations of the study were its cross-sectional design, the small sample size, the wide age range of the patient population, and the range of learning disability by which the children and young people were affected. Due to the paucity of teenage patients in the UK, individuals included in the 'over-12 years' age group included those aged up to 25 years. This was done so that the group size was large enough to

afford reasonable statistical analysis. Sample size was representative of this patient population in the UK however, as the majority of patients participated in the study. This must therefore be regarded as a strength of the study. Though the size of this patient sample was naturally small, it was reduced further for some of the analyses due to age parameters on some of the measures utilised, individuals' ability to complete the self-report measures due to learning disability, and the number of fathers that participated. In some instances individuals were excluded from analysis as their fathers had not participated. It is for these reasons that borderline relationships in the multiple regression analyses were discussed, as these relationships could indicate trends that, with a larger sample size, could show more significant relationships. Thus the author is aware that the sample size is small for the multiple regression analyses conducted, and the results are interpreted with caution, but feel this analysis to be useful to future research with this patient population.

The strengths of the study include its mixed methods design, which allowed for the collection of extensive data, both qualitative and quantitative. This has greatly enhanced our understanding of the experiences of this patient group and their families. Since this is a novel area of research the qualitative phase allowed the exploration of parents' experiences and the collection of rich data, which not only provided information regarding the experiences of having a child affected by MPS IH post-BMT, it also informed the design of the second phase of the study. In this subsequent phase, data was collected with the use of a multitude of validated measures. This included the collection of patient psychosocial, developmental and cognitive functioning data, and data relating to both mothers' and fathers' adjustment. Data collected therefore derived from parent- as well as patient self-report. This was enhanced by the development of a number of new measures which tapped into constructs pertinent to this patient group and their families, which were highlighted in the qualitative phase of this study. It is noted however, that external validity may have been challenged by the use of new measures that included items that were specific to characteristics of the MPS disorder and the BMT. Furthermore, though many of the new measures were piloted with parents of both healthy children and children affected by other MPS diseases, they perhaps lacked the reliability of other standardised measures.

The purpose of designing so many new measures was however to develop an in-depth understanding of the impact this condition has on the individual and their family and to identify risk and resistance factors *specific* to this patient group. These new measures would therefore benefit from further development with wider samples of families affected by MPS and related diseases, and from being refined into an MPS-specific quality of life measure. While there are commonalities in the psychosocial sequelae of chronic illness, there are also differences, and rare diseases such as MPS can pose unique challenges for individuals and their families. The development of a disease-specific quality of life measure would allow the regular, efficient, and effective assessment of psychosocial well-being at the practitioner level. This in turn would enable specialist guidance, intervention, and support to be given to individuals and families. Furthermore, knowledge and understanding about the condition may be enhanced and family stress, anxiety, and feelings of isolation reduced. It is important that such a measure includes objective measures of physical functioning and severity also, and that medical data is collected independently of the parents. A further factor that may have imposed some limitation on this study was the use of subjective data on medical outcomes and the lack of objective data on patient function and severity. It is possible that parental judgments in this respect were influenced by personal appraisals of their individual situation and the physical and emotional impact that this situation has had on them. It might have been more reliable to have obtained objective data on medical outcomes from hospital records and to have conducted independent measures of physical functioning. However, resource and time restrictions did not allow this.

The disability stress-coping model provided a useful framework from which to examine the psychosocial outcomes of bone marrow transplant for individuals affected by MPS IH. It allowed for the systematic examination of parent adjustment and of the contributors to adjustment and stress processing outcomes within a risk-resistance framework. It facilitated the examination of those outcomes, alongside other relevant variables, in relation to child adjustment outcomes. In the analysis, the variables worked well in the context of the model. However, the study would have benefited from the inclusion of a parent intrapersonal variable pertaining to self-efficacy, and from the variables being entered into the regression analyses in

theoretical groups that adhered more stringently to the model. Nonetheless, the findings highlighted the moderating effect of resistance factors on the relationship between illness-related stressors and parent and patient adjustment as predicted by the disability stress-coping model. The findings also indicate how parent stress processing factors function as stress processing factors relevant to child adjustment, and how social-ecological and illness-related risk factors impact directly on both parent and patient adjustment.

## **8.2. The Study's Contribution to Knowledge**

The findings of this study provide unique data on a previously unexplored area of research with a group of individuals affected by a rare and disabling chronic condition and their families. MPS IH is a rare genetic disorder of metabolism which is progressive and results in death within the first decade of life if untreated. It is routinely treated with bone marrow transplant in infancy but continues to present with progressive bone and joint disease post-BMT. Individuals affected by this condition are therefore of short stature, experience severe orthopaedic disability, and cognitive and adaptive functional impairment post-BMT. Research in the medical field has reported on the presenting features and diagnosis of MPS IH, and on BMT outcomes in terms of survival, adaptive and cognitive function. It has also reported on clinical trials of other treatment options like enzyme replacement therapy. This study explores the psychosocial development of this patient group post-BMT, which is an area of study that has previously been neglected.

The findings provide rich data which describe the experiences of families of children affected by this disorder. These highlight the rewards and challenges experienced by parents and increase understanding of some of the issues that individuals affected by the condition may face. They also illustrate the factors that contribute to both parent and patient adjustment. These findings enable the furtherance of research with this patient group and their families, and may be relevant to the study of other childhood chronic conditions. They also provide essential information with regard to patient adjustment outcomes and contributors to those outcomes, which can be practically used by health professionals, support organisations, and educators and applied to the

design of initiatives and intervention programmes which can support patients and the families and help improve outcomes. This research is therefore not only of academic importance in terms of the advancement of understanding regarding outcomes of BMT for this patient group, it is also of practical importance to the improvement of these outcomes.



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